

Spina Bifida Guideline

Quality of life: Guidelines for the care of people with spina bifida

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Abstract. Quality of Life (QOL) and Health-Related Quality of Life (HRQOL) are important concepts across the life span for those with spina bifida (SB). This article discusses the SB Quality of Life Healthcare Guidelines from the 2018 Spina Bifida Association's Fourth Edition of the Guidelines for the Care of People with Spina Bifida. The focus of these QOL Guidelines was to summarize the evidence and expert opinions on how to mitigate factors that negatively impact QOL/HRQOL or enhance the factors positively related to QOL/HRQOL, the measurement of QOL/HRQOL and the gaps that need to be addressed in future research.

Keywords: Myelomeningocele, spina bifida, pain, continence, quality of life, health-related quality of life, neural tube defects

1. Introduction

Quality of Life is defined as “an individual's perception of their position in life in the context of the culture and value systems in which they live and in relation to their goals, expectations and concerns” [1,2]. Health Related Quality of Life (HRQOL) is considered a sub-domain of Quality of Life (QOL) and measures a subjective perception of the impact of a health condition and/or its treatment on the individual [3–6]. HRQOL is most often multidimensional encompassing physical, emotional, social, and cognitive/occupational sta-

tus. The focus of the Quality of Life Spina Bifida (SB) Guideline was to summarize the evidence and expert opinions on how to mitigate factors that negatively impact QOL/HRQOL and enhance the factors positively related to QOL/HRQOL.

The measurement and study of QOL and HRQOL in SB are early in development. The rigor of most studies addressing these concepts is limited by: (a) sample size, diversity and response rate of participants, (b) measures that may not capture all domains of HRQOL and (c) measures that lack sensitivity to capture changes in QOL or HRQOL or (d) those which fail to reflect cultural differences [3,7]. Thus, the evidence that follows is preliminary and may be incomplete but summarizes the current state of the literature at the time that these guidelines were created.

QOL/HRQOL should be measured by condition and age-related instruments. The perceptions of both the

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parent and child/adolescent should be measured and the child's perception valued [3,7]. Parent report is often but not always lower than child/adolescent report [8,9]; children as young as eight can report on their QOL/HRQOL [10]. Use of HRQOL measures has been found useful in other chronic health conditions [7,11]. New age and SB-specific HRQOL instruments have been recently created (QUALAS-C, QUALAS-T, QUALAS-A) but have not been used extensively [8,9]. If time is limited, the adolescent self-report should preferentially be used over parent report.

When deciding on an instrument to use to measure QOL/HRQOL, it should be understood that some QOL measures and most HRQOL measures equate the ability to function to QOL/HRQOL such that any individual with a disability will have, by nature of the questionnaire, lower HRQOL than peers without disabilities. This conceptual equation devalues the lives of people with disabilities by automatically declaring that a person with a disability cannot have as good a quality of life as someone without disabilities. Measures that capture the individual's perception of how their condition (i.e., spina bifida) impacts their life are preferred [12]. The World Health Organization Quality of Life (WHO-QOL) Brief avoids measuring function by using items addressing perceived energy to do physical activities important to the individual [13]. Similarly, new SB and age specific measures address perception (e.g. bother, worry), not function [8,9].

Findings regarding the impact of SB on other domains of QOL/HRQOL (social, emotional, cognitive/school/work) for children, adolescents and adults are inconsistent [3], although one review of qualitative studies indicated more issues in the psychosocial domain of QOL than physical domain [6,14]. Evidence indicates most SB factors (e.g. level of lesion, severity of SB, ambulation) have generally had no or small associations with youth report QOL/HRQOL and only a modest relationship to parent report of generic QOL [3,7,15–18]. However, pressure ulcers and bladder infections were related to HRQOL in one study [19]. These relationships were most often seen in function-based instruments. In adults, there was some evidence that level of lesion, full time wheelchair use, latex allergy and hydrocephalus were associated with reduced HRQOL [19–25]. However, pain has consistently been related to reduced HRQOL in all ages, by both parent and self-report and across varied instruments [7,26].

Evidence consistently supports that bowel incontinence is associated with lower HRQOL and satisfaction with a bowel program is associated with

higher HRQOL [16,25,27]. Data on the relationship of bladder incontinence to QOL in children is inconsistent, but studies of adolescents and adults report that support for urinary continence contributes to overall HRQOL [28–31]. Using a new instrument (QUALAS-A) that specifically measures the impact of continence on adult HRQOL [8], any bowel continence and the amount, but not frequency, of urinary incontinence were related to the “Bladder and Bowel HRQOL subscale” but not to the “Health/Relationship or Esteem/Sexuality HRQOL subscales” [32]. There is little literature on sexuality and QOL, and using generic measures there was no relationship [33]. In studies to date, scoliosis status [34,35] has not been related to HRQOL. Only one study found obesity related to HRQOL in SB where as several found no relationship [36–38]. In contrast, obesity was related to HRQOL in typically developing children and those with other chronic health conditions [24,36–38].

Variables such as resilience (e.g., attitude towards SB, hope and future expectations, coping skills) have been strongly related to higher HRQOL and QOL [16, 17,39]. In contrast, depression, a lack of optimism, and reduced executive functioning were related to lower QOL/HRQOL [13]. Similarly, family variables such as higher family satisfaction and family resources have been related to higher QOL for adolescents and those over 18 years of age [13,16,17]. In order to foster QOL/HRQOL clinicians should develop strategies to optimize psychosocial well-being, bowel and bladder continence, and minimize the impact of pain, if present.

QOL or HRQOL should not be measured in isolation, and there may be components of HRQOL that are not measured by current instruments. If clinicians are going to address QOL they also need to address the factors important to the individual with SB and their family. An emerging concept, Family QOL (FQOL) may have usefulness in the care of individuals and families with SB [40,41].

2. Guidelines goals and outcomes

The goals of the QOL/HRQOL were both practical and aspirational

The aspirational objective of these guidelines was:

- Improve QOL across the lifespan in individuals with SB.

The secondary outcomes (specific goals) included:

- Increase QOL assessments in clinical practice.

Table 1
Questions that Informed the quality of life guidelines

Age group	Clinical questions
0–11 months	What factors are related to QOL?
1–2 years 11 months	What factors are related to QOL?
3–5 years 11 months	What factors are related to QOL?
6–12 years 11 months	What measures of QOL/HRQOL are the most efficient and useful?
	What factors are related to QOL?
	What might QOL assessment and improvement activities look like in clinical practice?
13–17 years 11 months	What measures of QOL and HRQOL are the most efficient and useful?
	What factors are related to QOL?
	What might QOL assessment and improvement activities look like in clinical practice?
18+ years	What measures of QOL and HRQOL are the most efficient and useful?
	What factors are related to QOL?
	What might QOL assessment and improvement activities look like in clinical practice?
	What measures of QOL and HRQOL are the most efficient and useful?

- Provide health care professionals with a better understanding of QOL and HRQOL measurement, potential issues related to available tools or tool development, and other factors related to QOL or HRQOL.

The ultimate goal to address the aspirational objective was:

- Clinicians of every specialty will integrate assessment of QOL and intervention to address QOL into clinical practice.

3. Methods

The methods for the review of the literature and development of the recommendations were designed by the executive committee of the SB guidelines work group [42]. In addition, because the search for QOL articles was inadvertently omitted from the central search process, the first author conducted a search of three databases, PubMed, CINAHL, and Psycinfo, 2000–2017 using the terms “spina bifida (myelomeningocele) and quality of life” and spina bifida (myelomeningocele) and health-related quality of life”. Forty-two articles, including three reviews, were identified that addressed these concepts. References of these publications were searched for any earlier overall QOL or HRQOL studies and six additional studies were added. In addition, select studies addressing emerging instruments were added as references. Along with expert opinion, this evidence informed the guidelines. In addition, the work group summarized the SB-specific measures HRQOL, generic QOL and HRQOL instruments, and family QOL instruments. Clinical questions were created to guide the organization of the guidelines (Table 1). The results of the 48 articles that addressed these concepts along with expert opinion informed the guidelines (Table 2).

4. Results

The quality of the studies reviewed for the guidelines were limited by the study designs, breadth of the studies reviewed and the limited variables considered and thus the recommendations need to be seen as preliminary (Table 2). There was some evidence that ambulation and LOL had small relationships to these outcomes but the relationships were most often to physical domains in instruments that measured function such as ability to walk. There was consistent evidence for both pain and incontinence, especially bowel incontinence, as important factors in QOL and HRQOL outcomes. In addition, psychosocial protective factors were also strongly related to these outcomes. Strategies that support family functioning, assist children and families in their efforts to develop protective beliefs (hope, attitudes, future expectations, active coping strategies), and optimize peer relationships were reported.

In addition, as the guidelines include recommendations for use of QOL and HRQOL instruments, the work group summarized and assessed generic QOL instruments, generic HRQOL instruments, SB-specific HRQOL instruments, and family QOL instruments and their potential use in the population with SB (Table 3). The critique used criteria developed by Waters et al. [4] but was expanded to include instruments developed since their original 2009 publication.

5. Discussion

QOL and HRQOL are important concepts that health care providers working with individuals and their families need to address across the life span. Central to understanding QOL/HRQOL in individuals with SB is the accurate measurement of these important demographic,

Table 2
Quality of life guidelines

Age group	Guidelines	Evidence
0–11 months	1. Consider strategies to assess and strengthen family functioning, which can be of critical importance in QOL outcomes in children	Clinical consensus, Family Functioning Guidelines [46]
	2. Address constipation because long-term constipation impedes the development of an effective bowel program.	Clinical consensus, Bowel Guidelines [57]
1–2 years 11 months	1. Consider strategies to assess and strengthen family functioning, which can be of critical importance in QOL outcomes in children.	Clinical consensus, Family Functioning Guidelines [46]
	2. Address constipation because long-term constipation impedes the development of an effective bowel program.	Clinical consensus, Bowel Function and Care Guidelines [57]
3–5 years 11 months	1. Assist families in their efforts to facilitate the development of protective psychosocial behaviors (e.g. showing affection, bouncing back when things don't go the child's way, showing interest in learning new things). Encourage independence, praise for accomplishment, and provide opportunities for fun.	Clinical consensus, Mental Health Guidelines [58] Clinical consensus, Family Functioning Guidelines [46]
	2. Address assessment of executive function.	Clinical consensus, Neuropsychology Guidelines [59]
	3. Target strategies to optimize the child's bowel program because bowel incontinence is consistently related to HRQOL.	Clinical consensus, Bowel Function and Care Guidelines [57]
6–12 years 11 months	Psychosocial well-being	
	1. Assist families in their efforts to facilitate the development of protective beliefs (e.g. hope, optimism, attitudes, future expectations, active coping strategies) and behaviors such as showing affection, bouncing back when things don't go their way, showing interest in learning new things, handling negative situations, and establishing and maintaining friendships.	Evidence [3,7,16,17,24], Mental Health Guidelines [58]
	2. Consider strategies to optimize peer relationships.	Evidence [60], Mental Health Guidelines [58]
	3. Consider strategies to assess and strengthen family functioning, which can be of critical importance in QOL outcomes in children.	Evidence [13,16,17], Family Functioning Guidelines [46]
	4. Refer to community resources that enhance protective factors, such as sports, camps, scouts, and other community programs.	Self-Management and Independence Guideline [61]
	5. Address assessment of executive function.	Evidence [62], Neuropsychology Guidelines [59]
	Continence	
	1. Target strategies to optimize bowel program effectiveness as any bowel incontinence has the greatest negative impact on QOL.	Evidence [25,27,30], Bowel Function Care Guidelines [57]
	2. Assess both volume and frequency of urinary incontinence, as volume may be more distressing than frequency.	Evidence [32], Urology Guidelines [63]
	Pain	
1. Evaluate presence and characteristics of any pain experienced.	Evidence [7,13,19,64]	
2. Develop strategies to address pain and its impact on school, work, recreation, and social activities.	Clinical consensus	
Measurement of QOL		
1. Use a systematic approach to evaluating QOL/HRQOL.	Evidence [3–5,7]	
2. Consider using both self and parent-report instruments.	Evidence [3,7]	
3. If feasible, use Spina Bifida and age-specific HRQOLs instruments that measure perception (“concerned about,” “worried about,” “avoid”) and avoid the problem of focusing on function in the physical domain (walking long distances, climbing stairs, jumping) when assessing children with Spina Bifida.	Evidence [3,4,7,9,10,12,32]	
4. Omit any measure that captures the impact in the physical domain. Emotional, social, and school/cognitive domains in most perception-based instruments are useful.	Table 3	
5. Consider using a single-item QOL question such as “How would you rate your quality of life?” on a scale of 0–100 with 0 = poor and 100 = excellent?	Evidence [7,16,17] Table 3	

Table 2, continued

Age group	Guidelines	Evidence
13–17 years 11 months	<p>Psychosocial well-being</p> <ol style="list-style-type: none"> 1. Assist families in their efforts to facilitate the development of protective beliefs (e.g. hope, optimism, attitudes, future expectations, active coping strategies) and behaviors such as showing affection, bouncing back when things don't go their way, showing interest in learning new things, handling negative situations, and establishing and maintaining friendships. 2. Consider strategies to assess and strengthen family functioning, which can be of critical importance in QOL outcomes in children. 3. Consider strategies to optimize peer relationships. 4. Consider each individual's unique priorities in QOL. 5. Refer to community resources such as sports, camps, scouts and other community programs that enhance protective factors. 6. Address strategies to compensate for executive functioning challenges. <p>Continence/mobility</p> <ol style="list-style-type: none"> 1. Target strategies to optimize bowel program effectiveness as any bowel incontinence has the greatest negative impact on QOL. 2. Investigate the child's satisfaction with her or his bowel program. Address concerns that will help to optimize the program. 3. Assess both volume and frequency of urinary incontinence as volume may be more distressing than frequency. 4. Consider functional mobility options that optimize societal participation. <p>Pain</p> <ol style="list-style-type: none"> 1. Evaluate presence and characteristics of any pain experienced. 2. Develop strategies to address pain and its impact on school, work, recreation, and social activities. (clinical consensus). <p>Measurement</p> <ol style="list-style-type: none"> 1. Use a systematic approach to evaluating QOL/HRQOL. 2. Consider using both self and parent-report instruments. 3. Use the new Spina Bifida HRQOL instrument that measures perception ("concerned about," "worried about") and avoids the problem of focusing on function in the physical domain (walking long distances, climbing stairs, jumping) when assessing children with Spina Bifida. Omit any measure that captures the impact in the physical domain. Emotional, social, and school/cognitive domains in most perception-based instruments are useful. 4. Use an age- and condition-specific instrument to assess QOL/HRQOL. 5. Evaluate both the child's self-report and the parent report of QOL/HRQOL. If assessment time is limited choose self-report. 6. Consider using a single-item QOL question(s) with follow up assessment if needed. For example: <ul style="list-style-type: none"> – "How would you rate your quality of life?" – "What makes up QOL for you?" – "What do you think would make your QOL better?" 	<p>Evidence [3,7,16,17,24], Mental Health Guidelines [58]</p> <p>Evidence [13,16,17], Family Functioning Guidelines [46]</p> <p>Evidence [60]</p> <p>Clinical consensus, Mental Health Guidelines [58]</p> <p>Clinical consensus, Self-Management and Independence Guideline [61]</p> <p>Evidence [62], Neuropsychology Guidelines [59]</p> <p>Evidence [25,27,30]</p> <p>Evidence [16], Bowel Function and Care Guidelines [57]</p> <p>Evidence [32], Clinical consensus, Urology Guidelines [63]</p> <p>Clinical consensus, Mobility and Function Guidelines [65]</p> <p>Evidence [7,13,19,64]</p> <p>Clinical Consensus</p> <p>Evidence [3–5,7]</p> <p>Evidence [3,7]</p> <p>Evidence [4,7,43,53] Table 3</p> <p>Evidence [3,7,8,10] Table 3</p> <p>Evidence [3,7,10,43]</p> <p>Evidence [7,16,17] Table 3</p>
18+ years	<p>Psychosocial well-being</p> <ol style="list-style-type: none"> 1. Identify strategies or resources to facilitate the development of protective beliefs (e.g. hope, optimism, attitudes, future expectations, active coping strategies) and behaviors such as showing affection, bouncing back when things don't go their way, showing interest in learning new things, handling negative situations, and establishing and maintaining friendships 	<p>Evidence [3,7,16,17,24], Mental Health Guidelines [58]</p>

Table 2, continued

Age group	Guidelines	Evidence
	2. Explore satisfaction with relationships and their sexuality.	Evidence [47], Sexual Health and Education Guidelines [66]
	3. Consider strategies to optimize peer relationships.	Mental Health Guidelines [58]
	4. Consider the importance of each individual's QOL unique priorities.	Clinical consensus
	5. Refer to community resources such as sports, camps, community advocacy groups, universities with strong programs to support students with disabilities, and other community programs that enhance protective factors.	Self-Management and Independence Guidelines [61]
	6. Address strategies to compensate for executive functioning challenges.	Evidence [62], Neuropsychology Guidelines [59]
	7. Consider strategies to enhance self-management behaviors.	Evidence [67], Self-Management and Independence Guidelines [61]
	Continence/mobility	
	1. Target strategies to optimize bowel program effectiveness as any bowel incontinence has the greatest negative impact on QOL in adults, especially in social domains.	Evidence [25,27,30]
	2. Investigate the adult's satisfaction with her/his bowel program Address concerns to optimize program.	Evidence [16]
	3. Assess both volume and frequency of urinary incontinence in adults, as volume may be more distressing than frequency.	Evidence [32]
	4. Consider functional mobility options that optimize societal participation.	Evidence [21], Mobility and Function Guidelines [65]
	Pain	
	1. Evaluate the presence and characteristics of any pain experienced.	Evidence [7,13,19,64]
	2. Develop strategies to address pain and its impact on school, work, recreation, and social activities.	Clinical consensus
	Measurement	
	1. Use a systematic approach to evaluating QOL/HRQOL.	Evidence [3-5]
	2. Use an age-and condition-specific instrument to assess HRQOL. Instruments that measure perception ("concerned about," "worried about," "avoid") and avoid the problem of focusing on function in the physical domain (walking long distances, climbing stairs, jumping) are preferred. Omit any measure that captures the impact in the physical domain. Emotional, social, and school/cognitive domains in most perception-based instruments are useful.	Evidence [4,7,8,12] Table 3
	3. Instruments like the WHOQOL-BREF avoid this issue using questions such as "Do you have enough energy for everyday activities?" or "To what extent do you feel that physical pain prevents you from doing what you need to do?" Spina Bifida-and-adult-specific measures also assess perception and avoid this issue.	Evidence [1,2,8,20] Table 3
	4. Evaluate both the adult's self-report and the parent report of QOL/HRQOL. If assessment time is limited choose self-report of QOL/HRQOL.	Evidence [3,7,10]
	5. Consider using a single-item QOL question(s) with follow up assessment if needed. For example:	Evidence [7,16,17] Table 3
	– "How would you rate your quality of life?"	
	– "What makes up QOL for you?"	
	– "What do you think would make your QOL better?"	

clinical and protective variables. To date, several instruments used in the assessment of QOL and HRQOL have an inherent bias, measuring function and not perception. It is critical for individuals with SB that these concepts be understood and perception-based rather than function-based instruments used. The individuals' perceptions of the impact of the disability on their

QOL/HRQOL, not the disability itself, is the central issue. Historical instruments used in the SB population have other limitations such as ceiling effects; and lack of reliability and validity data [3,7]. Emerging instruments address these limitations and make it possible to measure self and parent reports of QOL or HRQOL. However, if only one perspective can be collected, the

Table 3
Summary and assessment of QOL/HRQOL/FQOL instruments

Name, authors	Short description age range	Sub-scales	Criteria for assessing QOL/HRQOL measures (see legend for definition of criteria)*					Comments and recommendations		
			HF/QOL	Fam	Focus	Opp	Self		# Items	R&V
PedsQL™ (child or parent self-report) [68] Each 20 items	Versions (age): – Child (5–12) – Adolescent (13–18) – Young adult (18+ years) Parent and child report.	Physical, Emotional, Social, Cognitive (school/work).	1	2	1	3	1	3	3	Do not use physical scale. Emotional, social and cognitive scales may be useful especially if comparing to typically developing youth. However, heavy focus on functioning. Strong psychometrics across many CHC and typically-developing peers. Long; may be useful if specific sub-scales are of interest. No data on sensitivity to change. Multiple items with floor and ceiling effects. Not used extensively in Spina Bifida. Cerebral palsy comparison indicated it was outperformed by other measures [4].
			Generic instruments							
CHQ Child (187 items) or parent (50 items) self-report [69]	Parent and child version developed by experts using literature and other instruments.	Behavior, bodily pain, general health, mental health, parent impact, emotional, physical functioning, parent impact time, emotional/behavioral role, physical and self-esteem. Physical and psycho-social summary scores.	1	1	2	1	2	3	2	Only child generic instrument rated as 3 in all categories by Waters et al. [4]. No known use to date in US in children with Spina Bifida [7]. Focus groups, cognitive interviews and pilot testing 52 and 27-item versions. Validated in 12 European countries using over 22,000 children. Supported with internal and test-retest reliability. Preferred generic scale for adults with SB. Physical scale: while assessing the impact of physical status on QOL does so with items that do not automatically disadvantage individuals with a specific mobility-related impairment. Reference period: Last 2 weeks. Positively-worded and flexible for all conditions. (e.g. "Do you have enough energy for everyday life"?) Available at: http://www.who.int/mental_health/media/en/76.pdf .
			Generic instruments							
KID SCREEN [70] 27, 10 and 57-item self-report versions available	Ages 8–18. Developed and used primarily in Europe.	5 domains: – Physical well-being – Psychological well-being – Support – Peers – Financial resources.	3	3	3	3	3	3	3	Only child generic instrument rated as 3 in all categories by Waters et al. [4]. No known use to date in US in children with Spina Bifida [7]. Focus groups, cognitive interviews and pilot testing 52 and 27-item versions. Validated in 12 European countries using over 22,000 children. Supported with internal and test-retest reliability. Preferred generic scale for adults with SB. Physical scale: while assessing the impact of physical status on QOL does so with items that do not automatically disadvantage individuals with a specific mobility-related impairment. Reference period: Last 2 weeks. Positively-worded and flexible for all conditions. (e.g. "Do you have enough energy for everyday life"?) Available at: http://www.who.int/mental_health/media/en/76.pdf .
			Generic instruments							
WHOQOL-BRIEF The WHO QOL Group, 1998 [1] 26 items self-report adult instrument	Four domains. Shorter version of the original 100-item, 1997 instrument.	4 domains: – Physical health – Psycho-social health – Social – Environment 2 single items: – overall perception of QOL – overall perception of health.	3	3	2	3	3	2	3	Only child generic instrument rated as 3 in all categories by Waters et al. [4]. No known use to date in US in children with Spina Bifida [7]. Focus groups, cognitive interviews and pilot testing 52 and 27-item versions. Validated in 12 European countries using over 22,000 children. Supported with internal and test-retest reliability. Preferred generic scale for adults with SB. Physical scale: while assessing the impact of physical status on QOL does so with items that do not automatically disadvantage individuals with a specific mobility-related impairment. Reference period: Last 2 weeks. Positively-worded and flexible for all conditions. (e.g. "Do you have enough energy for everyday life"?) Available at: http://www.who.int/mental_health/media/en/76.pdf .
			Generic instruments							

Table 3, continued

Name, authors	Short description age range	Sub-scales	Criteria for assessing QOL/HRQOL measures (see legend for definition of criteria)*						Comments and recommendations	
			HF/QOL	Fam	Focus	Opp	Self	# Items		R&V
HRQOL-SB Parent (44 items) and adolescent (47 items) self-report [71]	Parent and adolescent versions: developed to assess QOL in SB.	No subscales identified.	3	3	2	3	3	2	1	Positively-phrased items. Many items with ceiling effects. Strong internal reliability. No factor structure, test-retest reliability, or sensitivity to change analyses. May be more appropriate for general assessment of younger child by parent. Only total score supported; no domain assessment possible. Use with caution. May be useful to assess HRQOL if previously used in a longitudinal study. Not a priority instrument for use in Spina Bifida. Good test-retest reliability, internal consistency, construct in both parent and child versions. Score 0–10. Parent minimally important difference is 0.10.
HOQ [72] Parent and child self-report Each 51 items	For children with hydrocephalus. Measures functional status.	Originated from focus groups. Subscales: Overall Health, Physical Health, Cognitive Health, Social Emotional Health.	1	2	1	1	3	2	1	
Quality of Life Assessments (QUALAS) with child, teen, and adult with spina bifida versions QUALAS Child (QUALAS-C) [9] 10 items	A family of three instruments created to evaluate living with Spina Bifida: child, teen, adult. Child (ages 8–12).	Reference: last 4 weeks. Responses are “never” to “always.” Five options plus alternative. Two scales: – Esteem/Independence – Bladder and Bowel.	3	3	3	3	2	3	2	Recommended for use with individuals who have SB. All three age versions: Based on qualitative and cognitive interviews. Strong input from families/those with Spina Bifida. Assessed using appropriately large enough samples to assess construct validity. All have good factor structure, strong internal reliability, test-retest reliability and minimally important difference (MID). The MID, useful in clinical practice, for child’s subscale “Esteem and Independence” and adult’s subscale “Health and Relationships” equal to or > 10. For all the other subscales in all 3 ages the MID is equal to or > 15.

Table 3, continued

Name, authors	Short description age range	Sub-scales	Criteria for assessing QOL/HRQOL measures (see legend for definition of criteria)*					Comments and recommendations		
			HF/QOL	Fam	Focus	Opp	Self		# Items	R&V
QUALAS-Teen (QUALAS-T) [43] 10 items	Teen Version (ages 13–17).	Two scales: – Family/Independence – Bladder and Bowel.	3	3	3	3	2	3	2	Some negative wording (“upset,” “embarrassed,” “bother you”) but generated from qualitative interviews and affirmed by cognitive interviews. No physical scale in child, teen or adult (could be an asset or liability). Child: Useful to assess how self-esteem and bowel and bladder status is perceived. See general critique of all age scales above. Typical question: “Did it annoy you if you could not do what other teenagers could do?” Besides “never” to “always” there is an alternative answer, “I could do what other teens do.” Teen: Useful to assess the two domains. May not be a useful measure of overall HRQOL.
QUALAS-Adult (QUALAS-A) [8] 15 items	Adult version (ages 18 and above) Positively-worded items In Health/Relationships and Esteem/Sexuality.	3 scales – Health/Relationships – Esteem/Sexuality – Bladder and Bowel	3	3	3	3	2	3	2	See general critique of all age scales above. Useful measure of domains assessed. May not be a useful measure of overall HRQOL. Important inclusion of items on sexuality (only instrument that does). Sexuality items might also be appropriate for older teens. Correlations between QUALAS-A and WHOQOL-BREF (overall QOL) were low except for high correlations with Health and Relationships domain ($0.63 \leq r \leq 0.71$), which supports the ability of the QUALAS-A to address HRQOL. Using Both instruments may be useful in understanding adult QOL and HRQOL. Bowel and Bladder scale same for teens and adults so can use same scale for those 13 or older.

Table 3, continued

Name, authors	Short description age range	Sub-scales	Criteria for assessing QOL/HRQOL measures (see legend for definition of criteria)*					Comments and recommendations		
			HF/QOL	Fam	Focus	Opp	Self		# Items	R&V
Spina Bifida Pediatric Quality of Life (SBPQ), a questionnaire for children with Spina Bifida [73]	Dutch scale developed by using existing items from other instruments (Ped-sQL and Fecal Incontinence QOL survey <i>n</i> = 10) and qualitative interviews yielded additional 25 items for total of 35 items for children 6–18 years mental age. Reduction during pretesting yielded final 30 item scale.	<ul style="list-style-type: none"> – General life – Physical functioning/daily activities – Social functioning, – Emotional functioning – School – Home – Hospital Includes questions on pain and energy (e.g. "Have you been too tired to do your regular activity?") Questions address last three months, 11 minutes to complete.	3	3	2	3	2	2	1	Only 62 patients used for initial assessment. Internal reliability good for most scales and ICC for stability. No factor analysis to confirm domains. Authors identify three items that "stand out" as negatively impacting QOL: – Feeling angry in the emotional domain (unclear whether this is related to SB) – the use of colon enemas in physical domain – missing activities as a result of doctors' visit/surgeries etc. Available in English but no data on English samples. Needs more psychometric evaluation before broad use.
QoL survey for people with a disability [74]. Self-report survey reduced to 30 items used for psychometric analysis	Goal was to develop a questionnaire to assess the impact of disability on QOL. Developed using 174 adolescents (age 15–24) with Spina Bifida, (38%); Muscular Dystrophy, and Fragile X syndrome to develop tool.	<ul style="list-style-type: none"> – Emotional health – Physical health – Independence – Activity limitation – Community participation 	3	1	3	3	1–2	1	1	Use with caution. Should avoid physical scale that addresses specific tasks (vigorous activities, running, and heavy lifting). Community participation scale may be useful. Activities scale would be more useful if stated in a positive manner (what the individual can do rather than focus on limitations). Instrument developed from other instruments. Preliminary psychometrics. No involvement of individuals or family members. Many items in article are useful and worth reviewing for those addressing transition.

Table 3, continued

Name, authors	Short description age range	Sub-scales	Criteria for assessing QOL/HRQOL measures (see legend for definition of criteria)*					Comments and recommendations	
			HF/QOL	Fam	Focus	Opp	Self		# Items
Single-item QOL [1,7,16,17] self-report used specifically with SB	A part of many instruments. Overall how would you rate your QOL?	Allows individual to determine domains important to them and prioritize domains based on personal perception.	3	1	3	3	3	2	Good for an overall perception; the person can give priority to their domains of importance. Does not help the health care provider identify what determines QOL for the individual. Would need a follow-up question to identify domains important to the individual. Some evidence of validity (related to variables as expected). Factor analysis and internal reliability – not applicable.
Fecal Incontinence (FIC) QOL survey not included as it addresses only one aspect of HRQOL. Refer to Sawin & Bellin, 2010 [7] for additional information.									
PedsQL™ Family Impact Model (parents self-report) [68]	Impact of pediatric CHC on parent's functioning. Family functioning subscale. One factor of general negative impact of pediatric CHC on social and familial systems.	Problems with physical, emotional, social, and cognitive functioning; communication; worry. Problems with family activities and relationships.	1	1	1	3	2	2	Some have used for FQOL. Authors indicate that it can be used for QOL assessment. Assesses impact on the family but may not be assessment of QOL. Heavy emphasis on function.
FQOL generic Tool [75] 3 item self-report instrument	Created for use with family with AYA with Spina Bifida. Parent and teen self-report 3 items: – How would you rate your QOL? – How would you rate your child/parent QOL? – How would you rate FQOL?? Rated from 0–100; summed and mean used.	Items allow responder to include domains important to them and to rank domains according to their own priorities. Rated from 0–100; summed and added.	3	2	3	3	3	1	The instrument has been evaluated in a sample of AYA with Spina Bifida [41] (n = 120), a comparison sample (n = 98) and parents of the AYA sample (n = instrument was found to have strong preliminary psychometrics including support for a single factor and high internal reliability).

Table 3, continued

Name, authors	Short description age range	Sub-scales	Criteria for assessing QOL/HRQOL measures (see legend for definition of criteria)*					Comments and recommendations		
			HF/QOL	Fam	Focus	Opp	Self		# Items	R&V
Beach Family QOL Scale (FQOL) [40,76]	Measures several aspects of perceived satisfaction. 5 domains: – Family Interaction – Parenting – Emotional well-being – Physical/material well-being – Disability-related support	25-item questionnaire; 5-point Likert-type response pattern. “Very dissatisfied” to “Very satisfied.” Available from Beach: https://www.midss.org/sites/default/files/fqol_survey.pdf	1	2	2	3	3	2	2	Widely used in the field of intellectual disabilities and families with children who have special needs. Developed at the Beach Center. Heavy emphasis on function. Detailed and long; may limit use in clinical practice.
<p>AYA = Adolescent/Young Adults; CHQ = Child Health Questionnaire; FQOL = Family Quality of Life; KIDSCREEN = [10,71,76] version; HOQ = Hydrocephalus Outcome Questionnaire; PedsQL = Pediatric Quality of Life family of tools; SR = Self-Report; QUALAS = Quality of life Assessments in SB for Child, Teen, Adult. *Criteria for Assessment of QOL/HRQOL [4]</p>										
Criteria Definition		Health/functioning = 1; midrange = 2; QOL = 3								
HF/QOL:	Original purpose of instrument	Low involvement of family = 1; midrange = 2; High involvement of family = 3								
Fam:	Origin of items	Functioning = 1; midrange = 2; well-being = 3								
Focus:	Actual focus of the instrument	No opportunity to self-report = 1; midrange = 2; self-report version available = 3								
Opp:	Opportunity to self-report	Negative wording = 1; midrange = 2; positive wording = 3								
Self:	Potential threat to self-esteem	Large number of items = 1; midrange = 2; small number of items = 3								
# Items:	Length	Poor or not demonstrated = 1; midrange = 2; excellent and demonstrated adequately = 3								
Reliability (R) & Validity (V)	Psychometric Properties									

For comprehensive assessment of generic QOL/HRQOL measures used in Spina Bifida see Bakamiene, et al., 2016; Sawin and Bellin, 2010, and Waters et al., 2009 [3,4,7].

individual's perception of their own QOL should have priority [3,7]. Critically important for all instruments are stability or test-retest reliability. Without this data, investigators cannot tell if changes in QOL/HRQOL measured by investigators are true changes or lack of reliability in the instrument. A set of age and SB-specific instruments that measure perception-based HRQOL have been developed and show promise for future use [8,9,43]. These child, teen, and adult instruments have established reliability and validity including test-retest reliability and minimally important differences (MID) for clinical use. The MID for child's subscale "Esteem and Independence" and adult's subscale "Health and Relationships" is equal to or > 10. For all the other subscales in all 3 ages the MID is equal to or > 15.

In addition the WHO-BRIEF, a generic instrument, offers a perception-based assessment of QOL for adults. The strengths and limitations of each of these instruments are addressed in Table 3. There was not enough experience with the concept or the tools to include FQOL in the guidelines but future investigation is warranted. Clinicians and researchers need to carefully review available instruments to determine which give them the information they are seeking.

Once measurement issues are addressed, better understanding the factors related to QOL/HRQOL becomes a priority. To date evidence would suggest that pain, specific continence issues, and select psychosocial variables are key [3,7]. Early interventions to address constipation and optimize bowel program effectiveness is important. Pain needs to be consistently assessed and addressed. Much of the evidence reviewed was identified from investigations of older children, teens, and adults. However, addressing the issues identified needs to begin early and be repeated often [44]. For example, addressing constipation and bowel continence early and developing an effective bowel program in toddlers can prevent bowel incontinence which has a major negative impact on child, teen and adult QOL/HRQOL. However, because of the multiple complex medical issues that arise early, many families and providers do not appreciate the priority of addressing a bowel program at a young age.

Evaluating modifiable and social determinants of health which predict HRQOL is critical for development of future interventions [45]. Providers in SB clinics need to increase interventions to enhance psychosocial protective factors and assist youth and their families to enhance social skills [3,7,18,24]. This need is particularly evident in adolescence and during the transition to

adulthood [13,18,46,47]. However, the building blocks for these skills are developed in earlier years. Innovative approaches to addressing these protective factors are needed as are evaluations of these approaches. In addition, during adolescence and adulthood, providers also need to consider the importance of each individual's QOL unique priorities. For many adults, this may include satisfaction with relationships and sexuality. Finally, while care of children, adolescents and some adults occurs in SB programs, most of the care for adults occurs in the broader community. It is critical that health care professionals in these settings assess the meaning of QOL for adults and initiate actions to address the identified issues. Providers in all settings need to be aware of the "disability paradox" and "response shifts" that can occur [48–50]. A recent qualitative study of adults with an implanted, destination therapy left ventricular assist device yielded the following conceptual definition: "I am able to live my life and do what I want, with some adjustments" [51]. Determining the meaning of 'living my life and doing what I want' needs to be a basic assessment parameter. Then professionals can assist *individuals* with spina bifida to develop adjustments they need to make to meet their goals.

The development of the guidelines also identified substantial research gaps. First, there is a need for continued refinement of HRQOL and QOL measurement including the relationships between individual and parent proxy reports. Continued research is needed to better identify the factors related to QOL/HRQOL and how change in these factors across time impacts QOL/HRQOL. Especially needed is research that extends the knowledge of what role factors such as finances, ethnic identity, religion and spirituality or aging play in QOL/HRQOL. Knowledge about QOL/HRQOL in the transition to adulthood and adult health care can advance understanding of this important time [52]. In addition, it is important to determine if measuring QOL/HRQOL in clinical practice actually leads to activities that improve QOL/HRQOL. Implementation research is needed to evaluate if emerging evidence on QOL/HRQOL is integrated into practice. If the emerging evidence is not being integrated into practice, there is a need to identify and address the barriers to implementing the findings. Since the publication of the guidelines, there has been emerging evidence which, in the view of the authors, is currently shaping the dialogue relevant to these guidelines. These findings will need to be integrated into ongoing care [53–56]. Finally, we need further research on the emerging concept of family QOL and its association with child outcomes.

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The Spina Bifida Association has already embarked on a systematic process for reviewing and updating the guidelines. Future guidelines updates will be made available as they are completed.

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Conflict of interest

Dr. Brei is the Medical Director of the Spina Bifida Association.

Dr. Sawin is a consultant to the Spina Bifida Association, on the Board of MetaStar (a quality improvement organization), and a funded investigator at the Wisconsin site, National Spina Bifida Patient Registry Project (funding from CDC).

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a solution proven effective for bowel care needs associated with spina bifida, spinal cord injury, traumatic brain injury, stroke, multiple sclerosis and general constipation in patients 12 years and older. A different formulation is available for children 2 to 12 years of age.



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Bowel continence significantly affects quality of life for individuals with spina bifida. Enemeez (docusate sodium) provides a fast, effective and safe bowel management program, reducing time spent on bowel care.

Neurogenic Bowel, Constipation & Fecal Incontinence in Spina Bifida Patients

The hallmark clinical presentation is the inability to “voluntarily” control the defecation process or the inability to routinely perform elimination of stool/waste from the body. Patients with Spina Bifida may also experience:

- Constipation, very often severe
- Bowel obstructions, rectal impaction with sensory loss
- Hemorrhoids
- Nausea and vomiting
- Abdominal pain, bloating-distention, cramping, and lethargy – “sluggish feeling”
- Diet changes = decreased appetite - “grazing-snacking”
- Dehydration = electrolyte disturbances and increased UTI risk
- Soiling and unplanned evacuation of stool / social anxiety

Enemeez® & Enemeez® Plus Mini-Enemas are a Fast, Effective & Safe Solution

- ✓ Fast, predictable results typically in 2-15 minutes.¹
- ✓ Can assist in reducing time spent with patient for dressing/redressing due to episodes of incontinence or fecal discharge.²
- ✓ Can virtually eliminate episodes of incontinence.³
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