

Guidelines for the Care of People with Spina Bifida

Quality of Life

Workgroup Members: Kathleen J. Sawin, PhD, CPNP-PC, FAAN (Chair); Timothy Brei, MD; Amy Houtrow, MD, PhD, MPH

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 status. The focus of this guideline is to mitigate the factors that negatively impact QOL/HRQOL and enhancing the factors positively related to QOL/HRQOL.

The measurement of QOL and HRQOL in Spina Bifida is early in its development. The quality of most studies are limited by sample size, diversity and response rate of participants, measures that may not capture all domains of HRQOL (e.g., impact of scoliosis and scoliosis repair on HRQOL) and measures the lack sensitivity to capture changes in QOL or HRQOL or 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, both the parent and child/adolescent perception should be measured and the child 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 Spina Bifida-specific HRQOL instruments have been recently created (QUALAS-C, QUALAS-T, QUALAS-A) (Appendix 1) but not been used extensively.⁸⁻⁹ If time is limited, the adolescent self-report should 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 (Appendix 1). 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 This focus on function is evident in the literature where QOL/HRQOL assessments of children with Spina Bifida are consistently lower in the physical domain as function (i.e. walking upstairs, running a distance) not perception, are measured.^{4,7}

Tools such as World Health Organization Quality of Life (WHOQOL) Brief uses items addressing perceived energy to do physical activities important to the individual and thus avoid this problem.¹³ Similarly, new Spina Bifida and age specific measures address perception (e.g. bother, worry), not function.⁸⁻⁹ Findings regarding the impact of Spina Bifida on other domains of

QOL/HRQOL for children, adolescents and adults (social, emotional, cognitive/school/work) are inconsistent, although one review of qualitative studies indicated more issues in the psychosocial domain of QOL than physical domain.^{3,6-7} Evidence regarding most Spina Bifida factors (e.g. level of lesion, severity of Spina Bifida, ambulation) have generally had no or small associations with youth report QOL/HRQOL and only a modest relationship to parent report of generic QOL.^{3,7,14-16} Pain has consistently been related in all ages, by both parent and self-report and across varied instruments.¹⁸

Other factors related to QOL/HRQOL found in recent literature include:

- Urinary tract infections and pressure injuries in children.
- Pressure injuries and latex allergy in adults.
- Level of lesion and hydrocephalus.
- Although Spina Bifida variables have been inconsistently related to QOL/HRQOL in children, some evidence indicates that level of lesion, full time wheelchair use, and hydrocephalus was associated with reduced HRQOL in adults.¹⁹⁻²⁴

Evidence consistently supports that bowel incontinence is associated with lower HRQOL and satisfaction with a bowel program is associated with higher HRQOL.^{15,24-25} 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.²⁶⁻²⁹

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.”³⁰ There is little literature on sexuality and QOL and using generic measures there was no relationship.³¹ In studies to date, scoliosis status³²⁻³³ has not been related to HRQOL. Only one study found obesity related to HRQOL in Spina Bifida.³⁴⁻³⁵ In contrast, obesity was related to HRQOL in typically developing children and those with other chronic health conditions.^{23,34-36}

Variables such as resilience (e.g., attitude towards Spina Bifida, hope and future expectations, coping skills) have been strongly related to higher HRQOL and QOL.^{15-16,37} 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.^{13,15-16} In order to foster QOL/HRQOL clinicians should develop strategies to optimize psychosocial wellbeing, bowel and bladder continence, and minimize the impact of pain, if present.

QOL or HRQOL should not be measured in isolation. 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 Spina Bifida and their family. An emerging concept, Family QOL (FQOL) may have usefulness in the care of individuals and families with Spina Bifida.³⁸⁻³⁹ FQOL has been measured with domain-specific instruments⁴⁰ and a generic FQOL tool (Appendix 1). There is not enough experience with the concept or the tools to include FQOL in the guidelines but future investigation is warranted.

Outcomes

Primary

1. Improve QOL across the lifespan in individuals with Spina Bifida.

Secondary

1. The information provided in this guideline gives the health care providers 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.
2. Increase QOL assessments in clinical practice.

Tertiary

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

0-11 months

Clinical Questions

1. What factors are related to QOL?

Guidelines

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)
2. Address constipation because long-term constipation impedes the development of an effective bowel program. (clinical consensus) (Bowel Function and Care Guidelines)

1-2 years 11 months

Clinical Questions

1. What factors are related to QOL?

Guidelines

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)
2. Address constipation because long-term constipation impedes the development of an effective bowel program. (clinical consensus) (Bowel Function and Care Guidelines)

3-5 years 11 months

Clinical Questions

1. What factors are related to QOL?
2. What measures of QOL/HRQOL are the most efficient and useful?

Guidelines

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) (Family Functioning Guidelines, Mental Health Guidelines)
2. Address assessment of executive function. (clinical consensus) (Neuropsychology Guidelines)

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)

6-12 years 11 months

Clinical Questions

1. What factors are related to QOL?
2. What might QOL assessment and improvement activities look like in clinical practice?
3. What measures of QOL and HRQOL are the most efficient and useful?

Guidelines

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.^{3,7,15-16,23} (Mental Health Guidelines)
2. Consider strategies to optimize peer relationships.⁴⁵ (Mental Health Guidelines)
3. Consider strategies to assess and strengthen family functioning, which can be of critical importance in QOL outcomes in children.^{13,15-16} (Family Functioning Guidelines)
4. Refer to community resources that enhance protective factors, such as sports, camps, scouts, and other community programs. (Self-Management and Independence Guidelines)
5. Address assessment of executive function.⁴¹ (Neuropsychology Guidelines)

Continence

1. Target strategies to optimize bowel program effectiveness as any bowel incontinence has the greatest negative impact on QOL.^{24-25,28} (Bowel Function and Care Guidelines)
2. Assess both volume and frequency of urinary incontinence, as volume may be more distressing than frequency.³⁰ (Urology Guidelines)

Pain

1. Evaluate presence and characteristics of any pain experienced.^{7,13,42-43}
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.^{4-5,7,44}
2. Consider using both self and parent-report instruments.^{3,7}
3. If feasible, use Spina Bifida and age-specific HRQOLs instruments that measure perception ("concerned about," "worried about," "avoid") 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.^{3-4,7,9-10,12} (Appendix 1)
4. Consider using a single-item QOL question^{7,15-16} such as "How would you rate your quality of life?" on a scale of 0-100 with 0=poor and 100=excellent? (Appendix 1) Individual and family factors associated with HRQOL in adolescents and young adults with Spina Bifida should be explored with follow up assessment if needed.

13-17 years 11 months

Clinical Questions

1. What factors are related to QOL?
2. What might QOL assessment and improvement activities look like in clinical practice?

3. What measures of QOL and HRQOL are the most efficient and useful?

Guidelines

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.^{3,7,15-16,23} (Mental Health Guidelines, especially the section on peer relationships)
2. Consider strategies to assess and strengthen family functioning, which can be of critical importance in QOL outcomes in children.^{13,15-16} (Family Functioning Guidelines)
3. Consider strategies to optimize peer relationships.⁴⁵ (Mental Health Guidelines)
4. Consider each individual's unique priorities important in QOL. (clinical consensus)
5. Refer to community resources such as sports, camps, scouts, and other community programs that enhance protective factors. (clinical consensus) (Self-Management and Independence Guidelines)
6. Address strategies to compensate for executive functioning challenges.⁴¹ (Neuropsychology Guidelines)

Continence/mobility

1. Target strategies to optimize bowel program effectiveness as any bowel incontinence has the greatest negative impact on QOL.^{24-25,28}
2. Investigate the child's satisfaction with her or his bowel program. Address concerns that will help to optimize program.¹⁵ (Bowel Function and Care Guidelines)
3. Assess both volume and frequency of urinary incontinence as volume may be more distressing than frequency.³⁰ (Urology Guidelines).
4. Consider functional mobility options that optimize societal participation. (clinical consensus) (Mobility Guidelines)

Pain

1. Evaluate presence and characteristics of any pain experienced.^{7,13,42-43}
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.^{3-5,7,44}
2. Consider using both self and parent-report instruments.^{3,7}
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.^{4,7,12} (Appendix 1)
4. Use an age- and condition-specific instrument to assess QOL/HRQOL.^{3,7-8,10} (Appendix 1)
5. Evaluate both the child's self-report and the parent report of QOL/HRQOL. If assessment time is limited choose self-report.^{3,7,10}
6. Consider using a single-item QOL question(s) with follow up assessment if needed.^{7,15,16} (Appendix 1). For example:
 - "How would you rate your quality of life?"
 - "What makes up QOL for you?"
 - "What do you think would make your QOL better?"

18+ years

Clinical Questions

1. What factors are related to QOL?
2. What might QOL assessment and improvement activities look like in clinical practice?
3. What measures of QOL and HRQOL are the most efficient and useful?

Guidelines

Psychosocial well-being

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.^{3,7,15-16,23} (clinical consensus) (Mental Health Guidelines, especially the section on peer relationships)
2. Explore satisfaction with relationships and their sexuality. (clinical consensus) (Sexual Health and Education Guidelines)
3. Consider strategies to optimize peer relationships. (clinical consensus) (Mental Health Guidelines)
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. (clinical consensus) (Self-Management and Independence Guidelines)
6. Address strategies to compensate for executive functioning challenges.⁴¹ (clinical consensus) (Neuropsychology Guidelines)
7. Consider strategies to enhance self-management behaviors.⁴⁷ (Self-Management and Independence Guidelines)

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.^{24-25,28}
2. Investigate the adult's satisfaction with her/his bowel program.¹⁵ Address concerns to optimize program.
3. Assess both volume and frequency of urinary incontinence in adults, as volume may be more distressing than frequency.³⁰
4. Consider functional mobility options that optimize societal participation.²⁰ (Mobility Guidelines)

Pain

1. Evaluate the presence and characteristics of any pain experienced.^{7,13,42-43}
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.^{4-5,7,44}
2. Consider using both self and parent-report instruments.^{3,7}
3. Use an age-and condition-specific instrument to assess HRQOL. Instruments that measures 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.^{4,7,12} (Appendix 1). Instruments like the WHOQOL-BREF (Appendix 1)^{1,2,19} 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.⁸

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.^{3,7,10}
5. Consider using a single-item QOL^{7,15-16} question(s) with follow up assessment if needed. (Appendix 1). For example:
 - "How would you rate your quality of life?"
 - "What makes up QOL for you?"
 - "What do you think would make your QOL better?"

Research Gaps

1. Need continued refinement of HRQOL and QOL measurement including the relationship of individual and parent proxy reports.
2. Continued research is needed to identify the factors related to QOL/HRQOL and how change in these factors across time impacts QOL/HRQOL. Especially needed is to extend the exploration of current factors to include whether finances, ethnic identity, religion and spirituality or aging with play a role in QOL/HRQOL.
3. Research is needed to determine if measuring QOL/HRQOL in clinical practice actually leads to activities that improve QOL/HQOL.
4. Research is needed to identify QOL/HRQOL during transition to adulthood and adult health care.⁴⁸
5. 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.
6. Need further research on the emerging concept of QOL in families and its association with child outcomes.

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Appendix 1: Summary and Assessment of QOL/HRQOL/FQOL Instruments

Summary and Assessment of QOL Instruments used in children, adolescents, and adults with Chronic Health Conditions (CHC) and their potential use in the population with Spina Bifida. The instrument uses criteria developed by Waters et al.⁴ and has been expanded to include additional instruments.

QOL/HRQOL Assessment Criteria Coding Table

1.	HF/QOL:	Original purpose of instrument	Health/functioning=1; midrange=2; QOL=3
2.	Fam:	Origin of items	Low involvement of family=1 midrange=2; High involvement of family=3
3.	Focus:	Actual focus of the instrument	Functioning=1; midrange=2; well-being=3
4.	Opp:	Opportunity to self-report	No opportunity to self-report=1 midrange=2; self-report version available=3
5.	Self-est:	Potential threat to self-esteem	Negative wording =1; midrange=2; positive wording=3
6.	# Items:	Length	Large number of items=1; midrange=2; small number of items=3
7.	Reliability & Validity	Psychometric Properties	Poor or not demonstrated=1; midrange=2; excellent and demonstrated adequately=3

Summary and Assessment of QOL/HRQOL/FQOL Instruments

Name, authors	Short description age range	Sub-scales	Criteria for assessing QOL/HRQOL measures							Comments and recommendations
			HF/QOL	Fam	Focus	Opp	Self-est	# Items	R & V	
Generic instruments										
PedsQL™ (Varni) (child or parent SR) ⁴⁹ 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

Name, authors	Short description age range	Sub-scales	Criteria for assessing QOL/HRQOL measures							Comments and recommendations
			HF/QOL	Fam	Focus	Opp	Self-est	# Items	R & V	
CHQ Child (N=187) or parent (N=50) SR ⁵⁰	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	Long; may be useful if specific subscales 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
KID SCREEN ⁵¹	Ages 8-18. 27, 10 and 57-item versions available.	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., 2009 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.
WHO QOL-BRIEF The WHO QOL Group, 1998 ¹	26 items in four domains. Shorter version of the 100-item, 1997 original instrument.	4 domains: -Physical health -Psycho-social health -Social - Environment 2 single items: -overall perception of QOL	3	3	2	3	3	2	3	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

Name, authors	Short description age range	Sub-scales	Criteria for assessing QOL/HRQOL measures							Comments and recommendations
			HF/QOL	Fam	Focus	Opp	Self-est	# Items	R & V	
		-overall perception of health								enough energy for everyday life"?) Available at: http://www.who.int/mental_health/media/en/76.pdf
Spina Bifida-Specific Instruments										
HRQOL-SB Parent and teen report ⁵²	2 versions: -child (parent report, 44 items) -adolescent (adolescent report, 47 items)		3	3	2	3	3	2	1	Positively-phrased items; many items with ceiling effect; 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.
HOQ ⁵³	For children with hydrocephalus. Measures functional status. No factor analysis.	Originated from focus groups	1	2	1	1	3	2	1	Not a priority instrument for use in Spina Bifida. Limited psychometric properties.
Quality of Life Assessments (QUALAS) with child, teen, and adult versions SR QUALAS Child (QUALAS-C) ⁹	A family of three instruments created to evaluate living with Spina Bifida: child, teen, adult. Child (ages 8-12), 10 items.	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. Good factor structure, internal and test-retest reliability.

Name, authors	Short description age range	Sub-scales	Criteria for assessing QOL/HRQOL measures							Comments and recommendations
			HF/QOL	Fam	Focus	Opp	Self-est	# Items	R & V	
QUALAS-Teen (QUALAS-T) ⁵⁴	Teen version (ages 13-17) 10 items	Two scales: -Family/Independence -Bladder and Bowel	3	3	3	3	2	3	2	<p>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."</p> <p>Some negative wording ("upset," "embarrassed," "bother you") but generated from qualitative interviews and affirmed by cognitive interviews.</p> <p>No physical scale in child, teen or adult (could be an asset or liability).</p> <p>Child: Useful to assess how self-esteem and bowel and bladder status is perceived.</p> <p>Teen: Useful to assess the two domains. May not be a useful measure of overall HRQOL.</p>
QUALAS-Adult (QUALAS-A) ⁸	Adult version (ages 18 and above) 15 items - all in health and relationships. Positively-worded.	3 scales -Health/Relationships -Esteem / Sexuality -Bladder and Bowel	3	3	3	3	2	3	2	<p>Useful measure of domains assessed.</p> <p>May not be a useful measure of overall HRQOL.</p> <p>Important inclusion on items on sexuality (only instrument that does). Sexuality items might be also appropriate for older teens.</p> <p>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 except for high correlations with Health and Relationships domain ($0.63 \leq r \leq 0.71$.) which supports the ability of the QUALAS-A.</p> <p>Bowel and Bladder scale same for teens and adults so can use same scale for those 13 or older</p>

Name, authors	Short description age range	Sub-scales	Criteria for assessing QOL/HRQOL measures							Comments and recommendations
			HF/QOL	Fam	Focus	Opp	Self-est	# Items	R & V	
Spina Bifida PQ QOL questionnaire for children with Spina Bifida ⁵⁵	<p>Dutch scale developed by using existing items from other instruments (PedsQL and Fecal Incontinence QOL survey n=10) and qualitative interviews.</p> <p>Yielded additional 25 items for total of 35 items for children 6-18 years mental age.</p> <p>Questions address last three months, 11 minutes to complete.</p>	<p>-Physical, social, and emotional function</p> <p>-School</p> <p>-Home</p> <p>-Hospital</p> <p>Child and parent versions available with picture book for children.</p> <p>Includes questions on pain and energy (e.g. "Have you been too tired to do your regular activity?")</p>	3	3	2	3	2	2	1	<p>Only 62 patients used for initial assessment.</p> <p>Internal reliability good for most scales and ICC for stability.</p> <p>No factor analysis to confirm domains.</p> <p>Authors identify three items that "stand out" as negatively impacting QOL:</p> <ul style="list-style-type: none"> -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. <p>Available in English but no data on English samples. Needs more psychometric evaluation before broad use.</p>
Development of a tool to describe overall health, social independence and activity limitation in AYA with disability ⁵⁶	<p>QOL tool for adolescents with a disability.</p> <p>Used with 174 adolescents with Spina Bifida, (38%); Muscular Dystrophy, and Fragile X syndrome to develop tool.</p>	<p>-Emotional health</p> <p>-Physical health</p> <p>- Independence</p> <p>-Activity limitation</p> <p>-Community participation</p>	3	1	3	3	1-2	1	1	<p>Use with caution.</p> <p>Should avoid physical scale that addresses specific tasks (vigorous activities, running, heavy lifting).</p> <p>Community participation scale may be useful.</p> <p>Activities scale would be more useful if stated in a positive manner (what the individual can do rather than focus on limitations).</p> <p>Instrument developed from other instruments. Preliminary psychometrics. No involvement of individuals or family members.</p> <p>Many items in article appendix are useful and worth reviewing for those addressing transition.</p>
Single-item QOL ^{1, 7, 15, 16}	<p>A part of many instruments.</p> <p>Overall how</p>	<p>Allows individual to determine</p>	3	1	3	3	3	3	2	<p>Good for an overall perception; the person</p>

Name, authors	Short description age range	Sub-scales	Criteria for assessing QOL/HRQOL measures							Comments and recommendations
			HF/QOL	Fam	Focus	Opp	Self-est	# Items	R & V	
	would you rate your QOL?	domains important to them and prioritize domains based on personal perception.								<p>can give priority to their domains of importance.</p> <p>Does not help the health care provider identify what determines QOL for the individual.</p> <p>Would need a follow-up question to identify domains important to the individual.</p> <p>Some evidence of validity (related to variables as expected).</p> <p>Factor analysis and internal reliability – not applicable.</p>
Fecal Incontinence (FIC) QOL survey not included as it addresses only one aspect of HRQOL. Refer to Sawin & Bellin, 2010 ⁷ for additional information.										
Family QOL (for full discussion of FQOL scales see HU et al, 2011)⁴⁴										
PedsQL™ Family Impact Model (parents SR) ⁴⁹	<p>Impact of pediatric CHC on parent's functioning.</p> <p>Family functioning subscale.</p> <p>One factor of general negative impact of pediatric CHC on social and familial systems.</p>	<p>Problems with physical, emotional, social, and cognitive functioning; communication; worry.</p> <p>Problems with family activities and relationships.</p>	1	1	1	3	2	2	2	<p>Some have used for FQOL. Authors indicate that it can be used for QOL assessment.</p> <p>Assesses impact on the family but may not be assessment of QOL.</p> <p>Heavy emphasis on function.</p>
FQOL generic tool ³⁹	<p>Created for use with family with AYA with Spina Bifida.</p> <p>Parent and teen self-report</p> <p>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</p>	<p>Items allow responder to include domains important to them and to rank domains according to their own priorities.</p> <p>Rated from 0-100; summed and added.</p>	3	2	3	3	3	3	1	The instrument has been evaluated in a sample of AYA with Spina Bifida (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.

Name, authors	Short description age range	Sub-scales	Criteria for assessing QOL/HRQOL measures							Comments and recommendations
			HF/QOL	Fam	Focus	Opp	Self-est	# Items	R & V	
	and mean used.									
Beach Family QOL Scale (FQOL) <small>38, 40</small>	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.

AYA = Adolescent/Young Adults; CHQ= Child Health Questionnaire; FQOL=Family Quality of Life. KIDSCREEN= (10,27,52 version); HoQ=Hydrocephalus Quality of Life; PedsQL = Pediatric Quality of Life—Varni family of tools; SR = Self-Report; QUALAS= Quality of life Assessments in SB for Child, Teen, Adult.

For comprehensive assessment of generic QOL/HRQOL measures used in Spina Bifida see Bakaniene, et al., 2016; Sawin & Bellin, 2010, and Waters et al., 2009.^{3,4,7}