

Guidelines for the Care of People with Spina Bifida

Self-Management and Independence

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Introduction

Self-management and independence are closely related constructs and are presented together. Self-management for youth and emerging adults with Spina Bifida is an active daily and flexible process in which youth and their parents share responsibility and decision-making for managing their condition, health, and well-being through a wide range of knowledge, attitudes, activities, and skills. The goal of this increasing responsibility is to develop the self-management behaviors needed to achieve independence and transition to adulthood and independent living.¹⁻² Self-management for all is the interaction of health behaviors and related processes that patients and families engage in to care for a chronic condition.³

Child autonomy provides a critical foundation for developing self-management and independence. For all children, autonomy begins early and is fostered by opportunities to make choices and to develop a sense of mastery. Most children with Spina Bifida achieve basic self-management and independence behaviors, (e.g., dressing appropriately, planning activities with peers, or cooking pre-planned meals) yet often lag 2-5 years behind their typically-developing peers in these behaviors.⁴ This gap may be due to the child's difficulties performing common everyday motor and processing activities in efficient and independent ways.⁵ Adaptation of performance and initiation of new steps may be especially challenging.⁵

Social skills in children are also important building blocks for independence. Many children with Spina Bifida need assistance with building adaptive social behaviors in peer interactions, specifically basic social skills such as reading social cues, clarity of thought and collaboration.⁶ Monitoring self-management learning is needed for all with cognitive functioning challenges, especially those with executive functioning, inattention and working memory issues.⁶⁻¹⁰ Educational programs in the home, school and broader community that offer opportunities to practice new behaviors are critical.

Youth do not enter adolescence with comprehensive knowledge of self-management (i.e. watching for signs of skin breakdown, bowel problems, shunt failure, and urinary tract infections), yet most develop this knowledge before age eighteen.^{1,4,11} Advanced self-management behaviors achieved by peers but not by individuals with Spina Bifida by age 18 are broad in scope and include doing their own laundry, cooking independently, managing their bank account without assistance, managing their allowance, and making their own appointments.⁴ It is not clear if these delays in skill development are developmentally appropriate for youths with Spina Bifida or due to a lack of expectations and support in the home, school, health system, or broader community. However, healthy family functioning was consistently related to better self-management outcomes across all developmental stages.^{9,11} Since there is evidence that responsibility in the home (e.g., chores and general decision-making) promotes self-management skill-building, individuals and families should be encouraged to expand their range of everyday living skills and responsibilities.^{4-5,11-14}

Potential self-management skill-building challenges identified from longitudinal research include older school-age children with Spina Bifida perceiving themselves as being more independent relative to parent assessment.¹⁵ Health care providers expect school-age and older children to perform self-management behaviors related to bladder programs, bowel programs, skin checks, and prevention of other secondary conditions.¹⁶ Yet, older children have reported that self-catheterization and bowel programs were a challenge, which often needed parent involvement.¹ In addition, both the family and child have had difficulty carrying out diet recommendations, bowel programs, and skin care.¹⁷ Thus, tailored interventions are needed to support growth in these areas.¹⁷

Children with Spina Bifida transitioning to adulthood are generally poorly prepared to self-manage their condition or live independently and enter young adulthood with preventable secondary conditions.¹⁸ Unhealthy behaviors continue into adulthood.¹⁹ Skin breakdown, along with fewer self-management behaviors, predicted hospitalization in this population.^{13,20} However, there is also evidence that improved self-management in young adults impacts health outcomes.

Adults were often without access to a usual source of health care or had gone without care due to barriers.²¹ Most adults over 18 years of age have not achieved optimal independence milestones in education, employment, and independent living.²²⁻²³ However, they were reported to have higher independence than those with other severe conditions.²⁴

Self-management interventions for youth with Spina Bifida and other Chronic Health Conditions (CHC) generally show at least one significant improvement, although a short workshop-based intervention for older children with Spina Bifida yielded no significant differences in groups.²⁵⁻²⁶ Family-oriented self-management interventions may be most effective in younger adolescents.²⁷ Camp-based psychosocial interventions promoting skills-development in goal-setting and problem-solving have shown promise in developmentally-diverse samples of children and adults with Spina Bifida.²⁸⁻²⁹ Other interventions using a problem-solving model combining education and home/community practice similarly improved child self-management skills and decreased parent burden. In addition, there is evidence that improved self-management in adults impacts health outcomes.³⁰ Rehabilitation interventions in young and middle-aged adults have improved all aspects of self-management and independence, with moderate- to large-effect sizes including self-efficacy, management of bowel and bladder incontinence, cognitive function, and psychosocial symptoms.^{13,31} Although tested mostly in adults, technology-based interventions hold promise for expanding self-management behaviors in youth as well.^{13-14,32-36}

Clinicians should consider using one of the valid and reliable generic or Spina Bifida-specific measures of self-management and independence.^{8,37-39} (Appendix A). Clinical assessment of the level of self-management and independence in those with Spina Bifida should specifically distinguish between the skills and behaviors the individual knows how to do and the behaviors they actually execute independently.⁴⁰ The evidence supports the need to have a structured, planned, and incremental approach to building self-management and independence skills beginning in early childhood, conveying expectations for developmentally-appropriate household responsibilities and increasingly involving the child in their care. Plans that accommodate cognitive learning styles or executive functioning status and purposefully, incrementally increase skills with multiple opportunities to practice new behaviors are central to achieve successful self-management and independence.

Outcomes

Primary

1. Perform effective self-management behaviors at the highest level of their abilities.
2. Achieve optimal independent living and employment, as well as maximal participation in society.
3. Young children develop autonomy, responsibility, and other foundational skills for self-management and independent living.

Secondary

1. Interventions that address the foundational skills necessary for complex self-management and independence behaviors are introduced throughout the lifespan, as appropriate.
2. Target foundational skills should include executive functioning skills, self-efficacy, self-regulation, and engaging in social activities.
3. Self-management and independence goals are evaluated yearly with the family, child, adolescent, and adult.

Tertiary

1. Adults with Spina Bifida over 18 who have a guardian responsible for their health care should perform self-management behaviors in the areas of medication management, prevention of complications, implementation of bladder and bowel programs, skin surveillance, and be able to communicate their findings to their guardian and/or health care providers at their highest level of ability.
2. Adults with Spina Bifida over 18 who do not need a guardian are fully responsible to self-manage their condition and independence (e.g., making appointments, ordering medications, arranging for transportation, conducting basic living skills like cooking and doing the laundry, managing money, managing insurance, and communicating with their health care provider).
3. Individuals with Spina Bifida interact effectively with family, health care providers, and others in the external environment in an independent manner.

0-11 months

Clinical Questions

1. What approaches optimize individual and family self-management and eventual independence?

Guidelines

1. Provide instruction and support to families regarding knowledge and skills needed to manage their child's Spina Bifida and related issues. (clinical consensus)
2. Provide orientation to families that include the expectation for eventual self-management and independence according to the individual's age and the status of their Spina Bifida. (Prenatal Counseling Guidelines)
3. Encourage families to expect participation in activities of daily life including tasks such as picking up toys, cleaning up, and imitative housework. (clinical consensus)
4. Evaluate and support family function. (Family Functioning Guidelines)
5. Identify and make referrals to early intervention programs. (clinical consensus) (Appendix: Early Intervention Services, Individualized Educational Plans (IEP) and 504 Plans)

1-2 years 11 months

Clinical Questions

1. What are the approaches that optimize individual and family self-management and eventual independence?

Guidelines

1. Provide instruction and support to families regarding knowledge and skills needed to manage their child's Spina Bifida and related issues. (clinical consensus)
2. Provide anticipatory guidance regarding developmental needs of children (such as exploration of environment, routines, and age-appropriate choices). (clinical consensus)
3. Teach families to offer daily age-appropriate choices such as choosing between two articles of clothes, two cereals for breakfast, and two books to read. (clinical consensus)
4. Encourage families to expect participation in daily life activities, including tasks such as picking up toys, cleaning up, and imitating housework. (clinical consensus)
5. Identify and make referrals to early education programs. (clinical consensus) (Appendix: Early Intervention Services, Individualized Educational Plans (IEP) and 504 Plans)

3-5 years 11 months

Clinical Questions

1. What approaches optimize independence and individual and family self-management in children with Spina Bifida?

Guidelines

1. Provide instruction and support to families regarding knowledge, skills, and behaviors needed to manage their child's Spina Bifida and related issues. (clinical consensus)
2. Discuss the need to expand the range of daily life activities and chores, as well as strategies to accommodate the child's learning style and/or mobility.^{41,42}
3. Provide anticipatory guidance that autonomy skills are maximized when positive behaviors are reinforced and clear and consistent consequences for inappropriate behavior are used. (clinical consensus) (Mental Health Guidelines, Neuropsychology Guidelines)
4. Refer to community resources such as early education programs that promote autonomy, self-efficacy, and other foundational independence skills. (clinical consensus) (Appendix: Early Intervention Services, Individualized Educational Plans (IEP) and 504 Plans)

6-12 years 11 months

Clinical Questions

1. What skills, abilities, and self-management behaviors should be targeted during age 6-12 years?
2. What are the most effective approaches to teach these skills and behaviors to children with Spina Bifida and their families?
3. Does specific skill training improve self-management behaviors (e.g., taking medication) and other independence behaviors?
4. What are optimal age expectations for specific self-management skills and behaviors (e.g. ability to self-catheterize; conduct skin checks; ability to describe their medication, its uses and side effects, and to take it on schedule; and describe their condition to a new professional) in children with Spina Bifida?
5. What instruments are available to measure self-management skills, abilities, and behaviors in children?

Guidelines

1. Provide instruction and support to children and families regarding the knowledge and skills needed to manage Spina Bifida and related independence issues. Teach the child basic self-management skills, including skills to prevent secondary conditions (clean

intermittent catheterization, skin care, equipment care, bowel and bladder care, wheel chair maintenance and propulsion) based on individual abilities. Focus on self-efficacy.⁴³ Children with Spina Bifida may develop foundational skills and self-management behaviors at a slightly later age (2-5 year delay) and may need more deliberate practice. However, most self-management behaviors are achievable by adults with Spina Bifida.^{4-5,12,17,43,45-46} (Neuropsychology Guidelines)

2. Assist families in learning how to incrementally involve the child in organizing school work and self-management activities and how to begin to transition from parents doing to child doing with parental oversight to eventually child doing without parent oversight.^{5,7,12,44}
3. Discuss the need to expand the range of daily life activities and chores as well as strategies to accommodate the child's learning style and/or mobility.^{4,7-8,11-12,14}
4. Serve as a resource to school systems regarding transportation, learning skills, health issues, and development of self-management skills. (clinical consensus)
5. Emphasize positive attitudes, self-esteem, assertiveness, self-efficacy and self-empowerment. (clinical consensus)
6. Assess peer relationships and encourage peer social involvement.^{1,6} (Mental Health Guidelines, Neuropsychology Guidelines)
7. Assess for potential patient, family, or environmental barriers to developing autonomy and independence, including family stress and conflict, and address in action plan.^{15,47} (Family Functioning Guidelines)
8. Assess bladder and bowel management programs for eventual independent self-management (Bowel Function and Care Guidelines, Urology Guidelines).
9. Consider using an age-and condition-appropriate assessment instrument (Appendix A) especially if the child has executive-functioning impairments.^{4,7-9,41-42}
10. Discuss with parents the need to help their child develop basic money management skills.⁴ If the child has an Individualized Educational Plan (IEP), encourage parents and the school to include money management skills in the child's IEP. (clinical consensus)
11. Encourage families to facilitate their child's language performance by creating intellectually- and culturally-enhancing activities in the child's typical environment.⁴⁸
12. Set beginning expectations for independent living. (clinical consensus)
13. Encourage use of technology to enhance self-management. (clinical consensus)

13-17 years 11 months

Clinical Questions

1. What approaches optimize individual and family self-management?
2. What skills, abilities and self-management behaviors should be targeted at age 13-17 years?
3. What are the most effective approaches to teaching these skills and behaviors to children age 13-17 years with Spina Bifida and their families?
4. Does specific skill training improve self-management behaviors (e.g., taking medication) and other independence behaviors?
5. What are optimal age expectations for specific self-management skills and behaviors (e.g. ability to self-catheterize; conduct skin checks; ability to describe their medication, its uses and side effects, take it on schedule, and describe their condition to a new professional) in children with Spina Bifida?
6. What instruments are available to measure self-management skills, abilities, and behaviors?

Guidelines

1. Evaluate self-management in appropriate areas (e.g. managing medications, prevention of complications, skin care, equipment care, bowel and bladder care, and making health

- care appointments). Assess self-efficacy⁴³ for these activities, considering that the child's ability to assume responsibility for health care encounters and other self-management of Spina Bifida is sequential. Full responsibility for self-management is critical for successful transition.^{4-5,12-13,17,45-46}
2. Assist families in knowing how to incrementally involve the child in organizing self-management activities and how to transition from parents doing to child doing with parental oversight to eventually child doing without parent oversight.^{5,7,12,44}
 3. Initiate a discussion and develop action plans to address deficits in self-management and independence skills, abilities, and behaviors, as needed.
 - Use a valid and reliable instrument to assess self-management skills, abilities, and performance of self-management and independence behaviors.^{8,41,49-50}
 - Encourage increasing responsibility for behaviors such as management of medication, bowel and bladder programs, and skin-monitoring.^{15,17} (Bowel Function and Care Guidelines, Integument (Skin) Guidelines, Urology Guidelines)
 - Support development of skills necessary for self-management (e.g., decision-making, goal-setting, self-regulation, and communication).^{3,11}
 - Evaluate and monitor cognitive functions as they underpin decision-making, goal-setting, self-regulation, self-management, socialization, and transition issues.⁷⁻¹⁰ (Neuropsychology Guidelines)
 - Assess the child's ability to use transportation. Encourage their enrollment in driver's education (adaptive, if needed) if the teen possesses the necessary cognitive and motor abilities. If driving is not realistic, teach (or encourage the family to teach) them how to use public transportation, van services for individuals with disabilities, or other transportation options.⁵¹ (clinical consensus)
 - Expand self-management interventions to encompass everyday living activities such as laundry, meal preparation, money management, managing finances, and making health care appointments.^{4,11,22,39,52}
 - Encourage the family to expand the range of responsibilities for daily life activities, chores, and jobs.¹¹
 - Evaluate the potential to eventually live independently (for those later in this age range) and connect them with housing resources (e.g. Centers for Independent Living). (clinical consensus)
 4. Encourage participation in IEP/504 planning that addresses self-management and transition skills. For those with an IEP, transition planning must be initiated by age 14. (Transition Guidelines)
 5. Support family functioning strengths related to self-management (navigating family stress, conflict, satisfaction, and family resources).^{47,49} (Family Functioning Guidelines)
 6. Involve the local Department of Vocational Rehabilitation and include vocational counseling in transition team planning. (clinical consensus)
 7. When it is developmentally appropriate, include time alone with the child to discuss self-management and independence topics as part of the visit. (clinical consensus)
 8. Discuss sexuality, contraception (including latex allergy precautions), marriage, childbearing issues, genetic counseling, and folic acid supplementation. (Latex and Latex Allergy in Spina Bifida, Men's Health Guidelines, Sexual Health and Education Guidelines, Women's Health Guidelines)
 9. Assess individual and system barriers to self-management and transition from pediatric to adult health care (e.g., responsibility for health management, advocacy, assertiveness, and insufficient adult services).^{21,52}
 10. Encourage the use of technology to enhance self-management.^{13,32-33,36,53}

11. Share expectations and resources for future independent living, transition to college or employment.^{18,44}
12. Provide consultation to adult providers with limited skill in providing care to those with congenital conditions such as Spina Bifida.⁵² (Transition Guidelines)

18+ years

Clinical Questions

1. What approaches optimize individual and family self-management?
2. What self-management skills, abilities, and behaviors lead to self-management and independent living in adults?
3. Does specific self-management skill training improve independence with self-management behaviors (e.g., taking medication and monitoring skin status)?
4. Is performing more self-management behaviors independently related to improved or positive health or functional outcomes (depression, quality of life, secondary conditions such as urinary tract infections, and pain)?
5. What health care and community supports optimize self-management, independence, and health outcomes?
6. Does increased independence with self-management increase community participation?
7. How can comprehensive preparation for self-management and independence be integrated into primary or specialty health care settings?
8. What instruments measure the individual's performance of self-management and independence behaviors in adulthood?

Guidelines

1. Evaluate full responsibility for implementing condition-specific self-management behaviors in appropriate areas, as needed (e.g. managing medications, preventing complications, monitoring skin care, maintaining equipment, bowel and bladder care, and ability to make health care appointments).^{4-5,12-13,17,41-42,45-46}
2. Reinforce the need for daily skin assessment, given the high incidence of skin breakdown on lower extremities (e.g. due to poor fitting leg braces) and risk for wound-related hospitalization.^{13,17,20} (Integument (Skin) Guidelines)
3. Evaluate if the adult has expanded self-management to encompass everyday living activities such as laundry, meal preparation, managing finances, making health care appointment and ordering supplies.
4. Initiate a discussion and develop an action plans to address deficits in self-management skills, abilities and behaviors, as needed. (clinical consensus)
 - Use a valid and reliable instrument to assess self-management skills, abilities and performance of self-management or independence behaviors over time in adults.^{8,41,49-50}
 - Support development of knowledge and skills necessary for self-management (e.g., self-efficacy, decision-making, goal setting, self-regulation, and communication).^{11,43}
 - Evaluate and monitor cognitive functions, as they underpin decision-making and self-management.^{1,18,45} (Neuropsychology Guidelines)
 - Assess the adult's ability to use transportation; encourage enrollment in driver's education (adaptive, if needed) if the adult possesses the necessary cognitive and motor abilities and has not done so already. If driving is not realistic, teach (or encourage the family to teach) the adult how to use transportation (e.g., public transportation, van services for individuals with disabilities, or other transportation options). (clinical consensus)

- Evaluate the young adult’s ability to live independently and connect with him or her with housing resources, such as Centers for Independent Living. (clinical consensus)
5. Encourage the use of technology in developing basic self-management skills. For instance, using email or a personal online health record, or patient portal to contact the clinic coordinator and physician with questions. Offer alternatives if this form of access is not available or appropriate.
 6. Encourage the use of technology programs to enhance self-management outcomes (e.g. using mobile health (mHealth) or telehealth tools to monitor skin breakdown or report response to medication for UTI).^{13,32-33,36,53}
 7. Expand the discussion of sexuality, contraception (including latex allergy precautions), marriage, childbearing issues, genetic counseling, and folic acid supplementation. (Sexual Health and Education Guidelines)
 8. Expand the discussion on child rearing and parenting issues and resources as appropriate. (clinical consensus)
 9. Discuss strategies for safe infant handling (e.g., holding an infant if you use a wheelchair or accessing a crib or car seat) with parents or expectant parents with mobility limitations. (clinical consensus)
 10. Encourage involvement in empowerment activities and organizations (e.g., sports, mentoring, camps, and local, national and international Spina Bifida, and other disability organizations).²⁸
 11. Support family functioning strengths related to self-management including family satisfaction and family resources.^{47,49} (Family Functioning Guidelines)
 12. Assess individual and system barriers to self-management (e.g., difficulties with self-advocacy, assertiveness, insufficient adult services).^{14-15,29,49}
 13. Refer to vocational rehabilitation, independent living centers, or other community agencies as appropriate. (clinical consensus)
 14. Provide information about accessible housing, financing, and appropriate outside agencies. (clinical consensus)
 15. Encourage planning and use of support services (e.g., in a college setting, services for students with disabilities) for self-management and independence in new environments. (Transition Guidelines)
 16. Encourage the use of wellness programs.³²
 17. Evaluate and support patients as their parents and caregivers age and assist individuals with Spina Bifida plan for changes in self-management and independence when their parents and caregivers will not be available. (clinical consensus)

Research Gaps

1. What are the foundational skills and abilities in young children that facilitate the development of self-management and independence behaviors in later childhood and adulthood?
2. What interventions optimize the development of these foundational skills and abilities early in childhood?
3. What are the barriers and facilitators for people with Spina Bifida to developing autonomy, self-management, and independent living skills?
4. What interventions are effective in closing the gap between self-management behaviors in individuals with Spina Bifida and their typically-developing peers? What interventions need to be targeted to patients and their parents to facilitate parental roles in self-management transitioning to coach and consultant as the patient nears adulthood?

5. Do interventions to enhance self-management and independence need to be delivered outside of clinical care?
6. What structure(s) of clinical services are optimal for coordinated, comprehensive transition to adult care?
7. Do routine clinical assessments of self-management behaviors, along with the development of action plans, in cooperation with the adolescent and their family yield improved outcomes?
8. What successful strategies can health care workers use to facilitate behaviors that encourage independence in children and adults with Spina Bifida and their parents?
9. What supports within the family and health care system lead to positive health and independence outcomes for children and adults with Spina Bifida?
10. How do the needs related to self-management and independence of young, middle-aged, and older adults change as they age with Spina Bifida?

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Appendix A. Self-Management Instruments

Name of instrument <i>Citation</i>	Short description: Number of items; age range; type of instrument; subscales	Psychometric evidence: Reliability and validity	Recommended use
Spina Bifida-Specific Instrument Developed with Samples of Youth with Spina Bifida			
<p>KKIS-SB Kennedy Krieger Independence Scales-Spina Bifida (KKIS-SB)</p> <p><i>The Kennedy Krieger Independence Scales-Spina Bifida Version: A Measure of Executive Components of Self-Management (KKIS-SB).⁸</i></p>	<ul style="list-style-type: none"> ● Twenty-two items. ● Caregiver-reported measure of self-care skills for individuals ages 10 and above. ● Based on assumption that self-care skills require adequate executive functioning and that other scales do not assess the executive burden of these tasks. ● Four response pattern options: <ul style="list-style-type: none"> ○ More than 90% of the time ○ 10-90% of the time ○ Less than 10% of the time ○ Not necessary or no opportunity ● Two subscales: Initiation of Routines And Prospective Memory. ● Initiation of Routines subscale items: <ul style="list-style-type: none"> ○ Keep room clean ○ Finish chores ○ Catheterize on time ○ Get out of bed on time ○ Hygiene on time ○ Take medication on time ● Prospective Memory subscale items: <ul style="list-style-type: none"> ○ Arrive at appointments on time ○ Arrange transportation ○ Look for skin breakdown ○ Start bowel program ○ Perform pressure relief ○ Write scheduled appointments 	<p>Evidence reported⁸</p> <ul style="list-style-type: none"> ● Psychometric analysis with a sample of 122 parents of individuals with Spina Bifida ages 10-29. <p>Reliability</p> <ul style="list-style-type: none"> ● Internal reliability $\alpha = 0.89$ ● Test-retest not reported <p>Validity</p> <ul style="list-style-type: none"> ● Exploratory factor analysis, reliability and construct validity using BRIEF (Behavior Report Inventory of Executive Function) were conducted. ● Factor analysis supported two subscales. ● Correlations between KISS-SB initiation of routines subscale and BRIEF summary scales ($r = -.031-0.56$) as well as five of the 8 BRIEF subscales (inhibit, shift, working memory and monitor) ($r = -.29$ to $-.62$) support validity of the KKIS-SB. ● Age-related changes and correlation with the Adaptive Behavior Assessment System scales also support validity of both KKIS-SB subscales 	<ul style="list-style-type: none"> ● Parent scales are recommended. ● Child report version exists but psychometric data have not been published. ● Contact developers for more information on child-report KKIS-SB. ● KKIS-SB provides a highly-specialized assessment of self-management abilities based on a known area of challenge in individuals with Spina Bifida (executive functioning).
<p>AMIS II (Interview)</p> <p><i>The Development of the Adolescent/Young Adult Self-Management and Independence Scale-AMIS II: Psychometric Data.³⁹</i></p>	<ul style="list-style-type: none"> ● AMIS II is a 17-item structured interview instrument that measures self-management behaviors in individuals ages 12 to adult. ● Parallel versions are available: <ul style="list-style-type: none"> ○ parent ○ adolescent/young adult/adult. ● These generic instruments have 3 questions (complication prevention, medication, and knowledge) that can be tailored to a specific condition. ● The individual is rated on how much of the behavior they actually perform and thus can be used as an outcomes measure. ● Response pattern: 7 options from 0%-100%. ● Two subscales: Condition Self-Management and Independent Living Self-Management. ● Condition Self-Management subscale: <ul style="list-style-type: none"> ○ condition knowledge ○ medication management ○ complication prevention 	<p>Evidence reported³⁹</p> <p>Initial psychometric analysis with a sample of 201 adolescents/young adults ages 12-25 with Spina Bifida and 129 of their parents.</p> <p>Reliability</p> <ul style="list-style-type: none"> ● Internal reliability $\alpha = .72-.89$. ● Test-retest intraclass correlation (ICC)=0.82 supporting test-retest <p>Validity</p> <ul style="list-style-type: none"> ● Factor analyses supported the two-factor AMIS II ● Validity supported by age-related changes and by moderate correlations with other related variables (parent-reported chores, responsibility and functional status and adolescent/young adult report of decision-making and functional status. $r = 0.30-0.61$) ● Validity also supported by use in published studies of transition-aged 	<ul style="list-style-type: none"> ● Parent and adolescent/young adult/adult versions are recommended. ● Scoring manual available from authors. ● Self-report version now available but no psychometric evidence to date ● Additionally, self-report instruments have been developed and are available for field testing. <ul style="list-style-type: none"> ○ AMIS II-SR-SB: 35-item Spina Bifida-specific instrument. ○ AMIS II-SR-G: 28-item generic instrument.

	<ul style="list-style-type: none"> ○ advocacy ○ family involvement ● Independent Living Self-Management subscale: <ul style="list-style-type: none"> ○ Making health care appointments ○ Ordering supplies ○ Household skills ○ Community living skills ○ Managing finances ○ Managing insurance 	<p>young adults. Increase in self-management over one year was related to decrease in depressive symptoms.</p>	
<p>Medical Self-Management and Transition Readiness</p> <p><i>Measurement of medical self-management and transition readiness among Canadian adolescents with special health care needs.⁵⁵</i></p>	<ul style="list-style-type: none"> ● Twenty-one-item measure of self-management and transition readiness for individuals ages 11-18 and their parents aimed at assessing awareness of their health care condition and ability to make decisions relative to health care. ● Response pattern: 1-5 Likert-type “strongly agree” to “strongly disagree” ● Item stems are primarily “know, can, understand.” For example: <ul style="list-style-type: none"> ○ “I know what medical insurance I have.” ○ “I can get myself to medical appointments.” ○ “I have discussed sexuality-related topics with my medical professionals.” ● Others are behaviors. For example: <ul style="list-style-type: none"> ○ “I take part.” ○ “I keep track.” ○ “I have discussed.” 	<p>Evidence reported: ⁵⁵</p> <ul style="list-style-type: none"> ● Psychometric study with a sample of 49 patients and their parents from a neurology clinic in Canada (only 1 person with SB) <p>Reliability</p> <ul style="list-style-type: none"> ● Internal reliability strong (0.89 to 0.93 adolescent/parent). ● No stability assessment. <p>Validity</p> <ul style="list-style-type: none"> ● Means, Standard Deviations (SD) and correlations to demographic and Scales of Independent Behavior (SIB). ● No factor analysis. ● Moderate correlations between adolescent/parent (r=0.56). ● Concurrent validity: <ul style="list-style-type: none"> ● Weak relationship to age in the parent report only. ● No relationship to parent education or transition program. ● Moderate relationship to a measure of skills needed to function at home, school, or work by adolescent and parent report. ● Major finding: medical self-management closely related to independence in other domains. 	<ul style="list-style-type: none"> ● Use with caution as no psychometrics established. ● Not as well developed as KKIS-SB or AMIS II.
<p>Spina Bifida Self-Management Profile (SBSMP)</p> <p>Sharing of Spina Bifida Responsibilities Scale (SOSBMR)</p> <p>Spina Bifida Independence Scales (SBIS)</p> <p><i>Condition self-management in pediatric spina bifida: A longitudinal investigation of</i></p>	<ul style="list-style-type: none"> ● A series of the instruments adapted from diabetes measures. Each measure collected from mother, father, and child. ● SBSMP: 14-item structured interview of adherence to treatments (diet, catheterization, bowel program, skin checks and exercise subscales). Indicates that the task is being completed but not by whom it was done. Items scored as either adherent or non-adherent (1,0). ● Sharing of Spina Bifida Responsibilities Scale (SOSBMR): 34 items showing who is primarily responsible for each task. Scoring (1=parent, 2= equal, or child=3). ● Spina Bifida Independence Scales (SBIS). Parent evaluation using 50 items of child’s knowledge and ability to do skills to manage Spina Bifida care 	<p>Evidence reported:</p> <ul style="list-style-type: none"> ● These measures reported in a study of 140 children with SB and their families. <p>Reliability</p> <ul style="list-style-type: none"> ● No internal reliability computed for SBSMP due to large number of “not applicable items.” Internal reliability for SOSBMR reported as Cronbach’s alpha>.60) for the 9 subscales. ● No-test-retest reliability reported for any of the scales. <p>Validity</p> <ul style="list-style-type: none"> ● Validity was generally supported by several relationships in the expected direction. ● Increased age was related to increased ability and responsibility. 	<ul style="list-style-type: none"> ● May have promise for future use, especially to compare to diabetes. ● Several scales are needed to measure these concepts and may have item burden in clinical practice.

<p><i>medical adherence, responsibility-sharing, and independence skills.</i>¹⁷</p>	<p>(yes, no, not sure, or NA) but does not measure if the child does them on a consistent basis.</p>	<ul style="list-style-type: none"> ● Increased ability was related to increased responsibility. ● However, the relationship between adherence and age was more variable. ● Although these scales have been used extensively in studies of children with diabetes, no formal assessment of validity in populations of children with Spina Bifida were reported. 	
<p>Generic Self-Management/Transition Readiness Instruments. No reported use in Spina Bifida.</p>			
<p>TRAQ (Transition Readiness Assessment Questionnaire)</p> <p><i>Measures the transition-readiness of youth with special health care needs.</i>⁵⁰</p>	<ul style="list-style-type: none"> ● Twenty-item scale created to reflect Stages of Change Theory (pre-contemplation, contemplation, preparation, action, maintenance) in individuals ages 12 years and older. ● Response Pattern: <ul style="list-style-type: none"> 1= I do not know how to do this 2 = I do not know how, but I want to learn 3 = I am learning how to do this 4 = I have started doing this 5 = I always do this when I need to ● Five subscales: <ul style="list-style-type: none"> ○ Managing medications ○ Appointment-keeping ○ Tracking health issues ○ Talking to providers ○ Managing daily activities 	<p>Evidence reported⁵⁰</p> <ul style="list-style-type: none"> ● Psychometric studies in three stages using 269, 178 and 526 participants respectively ranging in age from 12 to 26 years. <p>Reliability</p> <ul style="list-style-type: none"> ● Internal reliability strong=0.97 for total scale; 0.77-0.90 for subscales. ● No test-retest data reported. <p>Validity</p> <ul style="list-style-type: none"> ● Content validity: ethnographic interviews with adolescents/ family members to assess relevance, wording/ literacy level, intelligibility. ● Exploratory and Confirmatory Factor Analyses (RMSEA =0.23; GFI 0.92) support 5 subscales. ● All 5 subscales increase with age (p < 0.005). ● Gender differences found (females > males). 	<ul style="list-style-type: none"> ● Widely used and translated into multiple other languages. ● Excellent instrument for beginning discussions and facilitating movement to self-management. ● Does not measure increments of responsibility for implementing the behavior.
<p>UNC TR(x)ANSITION scale.</p> <p><i>A clinical tool to measure health care transition components from pediatric to adult care.</i>⁵⁷</p>	<ul style="list-style-type: none"> ● Thirty-three-item scale with 10 domains that uses a semi-structured interview format to measure issues in transition for those 12-20 years of age. ● Does not rely solely on patient report. Verified with information from the medical record. ● Ten domains: <ul style="list-style-type: none"> ○ Type of illness ○ Rx-Medications ○ Adherence ○ Nutrition ○ Self-management ○ Informed reproduction ○ Trade/school ○ Insurance ○ Ongoing support ○ New health providers 	<p>Evidence reported:⁵⁷</p> <ul style="list-style-type: none"> ● Pilot tested with a sample of 185 children/adults ages 12-20 with different chronic illnesses. <p>Reliability</p> <ul style="list-style-type: none"> ● Internal reliability supported by Item-total correlation scores (0.34 - 0.74). ● Inter-rater reliability was strong (kappa 0.71). ● No test-retest data reported. <p>Validity</p> <ul style="list-style-type: none"> ● Content and construct validity were satisfactory. ● Factor analysis not available ● Overall score was sensitive to advancing age (about a one-point increase in total score for each year of age). 	<ul style="list-style-type: none"> ● Promising generic tool. ● Each program should review items and determine if interview version is compatible with their clinic to determine use.

<p>PEDI-CAT <i>Pediatric Evaluation of Disability Inventory (PEDI) Computer Aided Test (CAT).</i>^{41, 42}</p>	<ul style="list-style-type: none"> ● PEDI-CAT for children and adults ages 0-21. Instrument uses Item-Response Theory to measure basic activities of daily living, mobility, social/cognitive and a new scale – responsibility by youth based on parent report. ● Responsibility scale has 51 item-bank. ● Five, 10 or 15 items are based on previous answers. ● Extension of the previous paper measure; addresses functional outcomes and adds responsibility ● Response pattern for Responsibility domain: <ul style="list-style-type: none"> ○ Adult/caregiver has full responsibility; the child does not take any responsibility. ○ Adult/caregiver has most responsibility and child takes a little responsibility. ○ Adult/caregiver and child share responsibility about equally. ○ Child has most responsibility with a little direction, supervision or guidance from an adult or caregiver. ○ Child takes full responsibility without any direction, supervision or guidance from an adult or caregiver. ○ I don't know. 	<p>Evidence reported:⁴¹</p> <ul style="list-style-type: none"> ● Psychometric assessment included a sample of parents of children and adolescents with disabilities (n=2205) as well as typically developing children (n=703). ● Only responsibility (self-management) scale data reported here. <p>Reliability</p> <ul style="list-style-type: none"> ● PEDI-CAT is more reliable and valid than the legacy (paper) PEDI measures. <p>Validity</p> <ul style="list-style-type: none"> ● Confirmatory factor analysis confirmed unidimensionality of the responsibility scale (CFI<.0.99, RMSEA=0.057). ● 5,10, or 15-item scales highly correlated with total item bank (r=>0.99). ● Paper version addressing functional outcomes has been used for children with Spina Bifida.⁵⁶ ● Additional validity testing needed on responsiveness and feasibility of use by parents with limited English. 	<ul style="list-style-type: none"> ● Excellent test of independence. One of few to span the 0-21 age group. ● Useful for measuring many daily and social foundational skills as well as the incremental performance of independence of behaviors. ● Recommended if clinic/program/organization has purchased technology and if technology available on routine basis. ● Theoretically could be used with young adults without intellectual disabilities as the reporter. ● Use of Item-Response Theory means a small number of items can tap a domain, e.g. responsibility. ● Limitation: Only validated with a parent reporter at the time this guideline was written.
<p>STARx Self-Management and Transition to Adulthood with Rx=Treatment Self-management and transition readiness assessment: development, reliability, and factor structure of the STARx questionnaire.⁵⁸</p>	<ul style="list-style-type: none"> ● Eighteen-item self-report survey for adolescents/ young adult (AYA) and parent report of three areas of transition readiness, disease knowledge, communication with medical provider, and self-management. ● Both paper and web-based administration versions available. ● Response pattern: 1-5 with “never” to “always” for behaviors; “nothing” to “a lot” for knowledge and “very hard” to “very easy” for self-management ● The instrument was developed in three phases including interviews of 29 AYA with a variety of chronic health conditions studies with strong input from AYA. <ul style="list-style-type: none"> ○ Subscales: <ul style="list-style-type: none"> ○ Medical Management ○ Provider Communication ○ Disease Knowledge ○ Adult Health Responsibilities ○ Resource Utilization 	<p>Evidence Reported:^{58, 59}</p> <ul style="list-style-type: none"> ● Initial psychometric assessment using sample of 194 AYA for reliability and factor structure after extensive item generation and pilot studies. Samples from 8 sites were for concurrent (n=267) and predictive validity (n=847). <p>Reliability</p> <ul style="list-style-type: none"> ● Internal reliability of total scale was strong (Cronbach alpha =0.80). Subscale reliabilities were moderate (α=0.44 to 0.77 with half below 0.70), ● Stability (n=26) was supported by ANOVCA analysis finding of no significant difference in two administrations of STARx. <p>Validity</p> <ul style="list-style-type: none"> ● Exploratory analysis yielded 6 factors: <ul style="list-style-type: none"> ○ 4 factors had 3 items. ○ 1 factor had 4 items (medication). ○ 1 factor had 2 items (resources). ○ Factor loadings were .31 to .88. 	<ul style="list-style-type: none"> ● Strong support for a brief measure of overall transition readiness. ● Recommended for self-report of AYA perceptions of knowledge, communication and select self-management behaviors in the last three months. ● Heavy emphasis on medication management.

		<ul style="list-style-type: none">• Concurrent validity supported by strong relationships to other transition measures (e.g. TRAQ $r=0.78$). Relationship of subscales of STARx to medication use, number of hospitalizations and length of hospitalizations support STARx predictive validity.	
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Resources:

The SBA “Beyond Crayons” resources are useful in developing the knowledge, self-efficacy, attitudes and skills necessary for self-management. They can be found here:

<http://spinabifidaassociation.org/beyond-crayons>