

Spina Bifida Guideline

Self-management and independence guidelines for the care of people with spina bifida

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Abstract. Self-management and independence behaviors are associated with improved health conditions common to spina bifida such as skin integrity and bowel and bladder management. While most children with spina bifida ultimately achieve basic self-care behaviors, (e.g., dressing appropriately, planning activities with peers, or cooking pre-planned meals), they often lag 2–5 years behind their typically-developing peers in these activities [1]. Valid and reliable condition-specific assessments of self-management and independence are critical to optimizing outcomes for this population. Partnerships among parents, clinicians, and youths with spina bifida are essential to implementing tailored interventions based on these assessments. The guidelines delineated in this article are informed by current self-management research for people with spina bifida and offer recommendations to promote self-management and independence across the lifespan.

Keywords: Myelomeningocele, spina bifida, self-management, independence, guidelines, neural tube defects

1. Introduction

Research suggests that more than 75% of individuals born with spina bifida [SB] will survive into adulthood [2] with evidence underscoring the critical importance of self-management in extending life expectancy. According to the *Guidelines for the Care of People with Spina Bifida* [3]:

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 [4,5].

Self-management may also be thought of as the interaction of health behaviors that affected individuals and families engage in to care for a chronic condition [6].

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Within the SB population, effective self-management is particularly important for preventing co-morbidities and secondary complications such as skin breakdown, renal dysfunction, and bladder and bowel incontinence [7,8]. Self-management is also central to a successful transition from adolescence into emerging adulthood, participating fully in society, and combating vulnerability, stigma, and discrimination. SB is a congenital condition; therefore the building blocks of self-management must begin early in childhood and in partnership with families and multi-disciplinary providers [9]. Therefore, these guidelines begin in infancy and progress through adulthood.

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 SB ultimately 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 [1]. This gap may be due to the child's difficulties performing common everyday motor and processing activities in efficient and independent ways [1]. Adaptation of performance and initiation of new steps may be especially challenging [10].

Social skills in children are also important building blocks for independence. Many children with SB need assistance building adaptive social behaviors in peer interactions, specifically basic social skills such as reading social cues, clarity of thought, and collaboration [10]. Monitoring self-management learning is needed for all with cognitive functioning challenges, especially those with executive functioning, inattention, and working memory issues [11–15]. Educational programs in the home, school, and broader community that offer opportunities to practice new behaviors are critical.

Youth with SB often do not enter adolescence with a comprehensive knowledge base of how to self-manage their condition (i.e. watching for signs of skin breakdown, bowel problems, shunt failure, and urinary tract infections) yet most develop this knowledge before age eighteen [1,4,16]. Advanced self-management behaviors for community living (i.e., managing a bank account, cooking independently) are typically achieved by peers without complex disabilities by age 18, but not as commonly by individuals with SB [1]. It is not clear if these difficulties reflect a delay in development for youth with SB or are due to a lack of expectations and

support in the home, school, health system, or broader community. However, research suggests healthy family functioning characterized by open communication and shared decision-making is related to more optimal self-management outcomes across all developmental stages [13,16].

Since there is evidence that responsibility in the home (e.g., chores and general decision-making) promotes self-management and skill-building, individuals and families should be encouraged to expand the range of everyday living skills and responsibilities for youth with SB [1,16–21]. These findings are complicated by longitudinal studies indicating that older school-age children with SB perceive themselves as being more independent relative to parent assessment [21]. Moreover, while 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 [22], older children have reported that self-catheterization and bowel programs were a challenge and often required parent involvement [4]. In addition, both the families and children have had difficulty carrying out diet recommendations, bowel programs, and skin care [23]. Thus, tailored interventions are needed to support growth in these areas [23].

Adolescents with SB transitioning to emerging adulthood are generally poorly prepared to self-manage their condition or live independently and are therefore at risk for preventable secondary conditions such as skin breakdown and UTIs [24]. This vulnerability appears to persist into adulthood [25]. Other research has identified associations between lower self-management behaviors and higher hospitalization rates in this population [7,26]. However, there is evidence that improved self-management in young adults is associated with decreased depressive symptomology [27]. In addition, there is evidence in other conditions such as asthma that increased self-management improves health outcomes and healthcare utilization [28–31]. Most adults with SB over 18 years of age have not achieved optimal independence milestones in education, employment, and independent living [32]. However, they were reported to have higher independence than those with other severe conditions such as fragile X syndrome or muscular dystrophy [33].

Self-management interventions for youth with SB and other chronic health conditions generally show significant improvement in at least one area, although a short workshop-based intervention for older children with SB yielded no significant differences in interven-

tion and control groups [34,35]. Family-oriented self-management interventions may be most effective in younger adolescents [36]. Camp-based psychosocial interventions promoting skills-development in goal-setting and problem-solving have shown promise in developmentally-diverse samples of children, adolescents, and adults with SB [11,37]. Another intervention using a problem-solving model combining education and home/community practice similarly improved perceived family stress associated with SB condition management but not youth self-management [36]. 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 [8,26]. Although tested mostly in adults, technology-based interventions hold promise for expanding self-management behaviors in youth as well [20,38–42].

1.1. Guidelines, goals and outcomes

The goals and desired outcomes of the Self-Management and Independence Guidelines were developed to be both practical and aspirational.

1.1.1. Primary outcomes

The aspirational objectives of these guidelines were to improve our developmental methodology to facilitate children and adults with spina bifida to:

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

1.1.2. Secondary outcomes

The guidelines address practical goals to achieve these aspirational objectives, including:

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

1.1.3. Tertiary outcomes

By supporting development of these important self-management and independence skills, the guidelines aim to promote independent behaviors and activities of adolescents and adults with SB, including:

- Adults over 18 with SB who have a guardian responsible for their health care perform self-management behaviors in the areas of medication management, prevention of complications, implementation of bladder and bowel programs, skin surveillance, and have the ability to communicate their findings to their guardians and/or health care providers at their highest level of ability.
- Adults over 18 with SB 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).
- Individuals with SB interact effectively with family, health care providers, and others in the external environment in an independent manner.

2. Methods

The methods for the review of the literature and development of the recommendations were designed by the Executive Committee of the SB Guidelines [43]. Independence had been a topic in previous guidelines, however self-management was added in this edition. The central staff conducted a search of the literature from 2006 to 2016 using the search terms “independence and spina bifida” or “independence and myelomeningocele”. This search yielded 18 studies that addressed independence. In addition, because the search for self-management studies was inadvertently omitted from the central search process, one author (Au 2) conducted a search of three databases, PubMed, CINAHL, and Psychinfo for the dates 2006–2016 using the terms “spina bifida and self-management or spina bifida and independence.” This search yielded an additional 25 studies. References of these publications were searched for any earlier self-management studies; three additional studies were added. In addition, later during the review process two studies were identified and included. Early in the literature review, it became apparent that the recommendations of the self-management and independence working groups were similar. Therefore,

Table 1
Clinical questions that informed the self-management and independence guidelines

Age group	Clinical questions
0–11 months	1. What approaches optimize individual and family self-management and eventual independence?
1–2 years	1. What approaches optimize individual and family self-management and eventual independence?
11 months	
3–5 years	1. What approaches optimize independence along with individual and family self-management in children with spina bifida?
11 months	
6–12 years	1. What skills, abilities, and self-management behaviors should be targeted during age 6–12 years?
11 months	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; describe their medication, its uses and side effects, and 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?
13–17 years	1. What approaches optimize individual and family self-management?
11 months	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; 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?
18+ years	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?

the decision was made to combine the two sections. The blended working group consisted of: a PhD social worker, a PhD physical therapist, a PhD nurse, a rehabilitation physician, and a developmental pediatrician. Forty-eight studies informed the combined recommendations. In addition to the age-specific recommendations, the combined work group created a table describing the SB self-management instruments identified in this search and seven additional generic self-management instruments with references supporting their reliability and validity. Clinical questions were created to guide the organization of the guidelines (Table 1). The results of the 48 studies that addressed these concepts along with expert opinion informed the guidelines.

3. Results

Evidence continues to mount in support of assessment and education of families and children/adolescents/young adults with SB around self-management is-

sues (Table 2). The importance of these issues in adulthood is undisputed. Using valid and reliable tools to measure these concepts is well-supported (Table 3). Evidence is emerging for the success of community-based educational and training programs to improve self-management. More research is needed to support and refine these programs. In addition, we do not yet know the most effective developmental steps that encourage independence from a young age in this population.

4. Discussion

There are several themes that have emerged from these guidelines. While it is important to understand the risk factors that complicate self-management such as level of the lesion, cognitive ability, and functional mobility, the trajectory of self-management skill development can be positively impacted independent of these complicating elements. Families and clinicians must work together to assess child readiness for self-

Table 2
Self-management guidelines

Age group	Guidelines	Evidence
0–11 months	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 [57]
	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 Family Functioning Guidelines [58]
	4. Evaluate and support family function.	Clinical consensus Appendix: Early Intervention Services, Individualized Educational Plans [IEP] and 504 Plans [59]
	5. Identify and make referrals to early intervention programs.	Clinical consensus Clinical consensus Appendix: Early Intervention Services, Individualized Educational Plans [IEP] and 504 Plans [59]
1–2 yrs 11 months	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 clothing, two cereals for breakfast, or 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 [59]
3–5 years 11 months	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.	Evidence [60,61]
	3. Provide anticipatory guidance so 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 [62], Neuropsychology Guidelines [63]
	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 [59]
6–12 years 11 months	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, wheelchair maintenance, and propulsion) based on individual abilities. Focus on self-efficacy. 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.	Evidence [1,16–18,23,64,65] Neuropsychology Guidelines [63]
	2. Assist families in learning how to incrementally involve the child in organizing schoolwork and self-management activities. Specifically, encourage transition to having the child complete these activities initially with parental oversight and eventually independently.	Evidence [12,17,18,38]
	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.	Evidence [4,10] Mental Health Guidelines [62], Neuropsychology Guidelines [63] Evidence [21,36] Family Functioning Guidelines [58]

Table 2, continued

Age group	Guidelines	Evidence	
6–12 years 11 months	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.	Evidence [4,10] Mental Health Guidelines [62]	
	7. Assess for potential patient, family, or environmental barriers to developing autonomy and independence (including family stress and conflict) and address in action plan.	Evidence [21,36,58]	
	8. Assess bladder and bowel management programs for eventual independent self-management.	Evidence [66]	
	9. Consider using an age- and condition-appropriate assessment instrument especially if the child has executive-functioning impairments (Table 3).	Evidence [1,12,15,59–61]	
	10. Discuss with parents the need to help their child develop basic money management skills [1]. If the child has an Individualized Educational Plan (IEP), encourage parents and the school to include money management skills in the child's IEP.	Evidence [1] 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.	Evidence [67]	
	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	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-efficacy for these activities, considering that the child's ability to assume responsibility for health care encounters and other self-management of spina bifida can progress. Full responsibility for self-management is critical for successful transition.	Evidence [1,17,18,23,38,64,65,68]
		2. Assist families in knowing how to incrementally involve the child in organizing self-management activities. Specifically, encourage transition to having the child complete these activities initially with parental oversight and eventually independently.	Evidence [12,17–19]
		3. Initiate a discussion and develop action plans to address deficits in self-management and independence 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 and independence behaviors (Table 3).		Evidence [13,32,46,60,69]	
– Encourage increasing responsibility for behaviors such as management of medication, bowel and bladder programs, and skin-monitoring.		Evidence [21,23,66,70,71]	
– Support development of skills necessary for self-management (e.g., decision-making, goal-setting, self-regulation, and communication).		Evidence [6,16]	
– Evaluate and monitor cognitive functions as they underpin decision-making, goal-setting, self-regulation, self-management, socialization, and transition issues.		Evidence [12–15] Neuropsychology Guidelines [63]	
– 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.		Evidence [72] 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.		Evidence [1,16,32,46,73]	
– Encourage the family to expand the range of responsibilities for daily life activities, chores, and jobs.		Evidence [16]	
– 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 [74]	
5. Support family functioning strengths related to self-management (navigating family stress, conflict, satisfaction, and family resources).		Evidence [36,75]	
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		

Table 2, continued

Age group	Guidelines	Evidence
13–17 years 11 months	8. Discuss sexuality, contraception (including latex allergy precautions), marriage, childbearing issues, genetic counseling, and folic acid supplementation.	Latex and Latex Allergy Guidelines [76] Men's Health Guidelines [77] Sexual Health and Education Guidelines [78] Women's Health Guidelines [79] Evidence [73,80]
	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).	
	10. Encourage the use of technology to enhance self-management.	Evidence [38–40,42,81]
	11. Share expectations and resources for future independent living, transition to college or employment.	Evidence [19,24]
	12. Provide consultation to adult providers with limited skill in providing care to those with congenital conditions such as spina bifida.	Evidence [73] Transition Guidelines [74]
18+ years	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).	Evidence [1,17,18,23,38,60,61,65,68]
	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.	Evidence [26,36,38] Integument {Skin} Guidelines [70]
	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.	Clinical consensus
	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 (Table 3).	Evidence [13,60,69,75]
	– Support development of knowledge and skills necessary for self-management (e.g., self-efficacy, decision-making, goal setting, self-regulation, and communication).	Evidence [16,64]
	– Evaluate and monitor cognitive functions, as they underpin decision-making and self-management.	Evidence [4,24,68] Neuropsychology Guidelines [63]
	– 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, 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.	Clinical consensus
	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].	Evidence [10,38–40,42,82]
	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 [78]
	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).	Evidence [11]
	11. Support family functioning strengths related to self-management including family satisfaction and family resources.	Evidence [58,75,83]

Table 2, continued

Age group	Guidelines	Evidence
18+ years	12. Assess individual and system barriers to self-management (e.g., difficulties with self-advocacy, assertiveness, and insufficient adult services).	Evidence [20,21,37,75]
	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.	Clinical consensus
	16. Encourage the use of wellness programs.	Evidence [39]
	17. Evaluate and support patients as their parents and caregivers age and assist individuals with spina bifida with planning for changes in self-management and independence when their parents and caregivers will not be available.	Clinical consensus

management and collaboratively cultivate those skills over time. Clinicians should consider using one of the valid and reliable generic or SB-specific measures of self-management and independence [13,44–46]. Clinical assessment of the level of self-management and independence in those with SB should specifically distinguish between the skills and behaviors the individual knows how to do and the behaviors they actually execute independently [47]. 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. While the science of self-management for individuals with SB is not completely established, the general health behavior literature supports the powerful effect of expectations and self-efficacy [48,49]. The expectation that children with spina bifida will grow into independent teenagers and adults must be supported by health care providers starting in infancy and increasing over time [50]. Typical developmentally appropriate expectations of putting away toys for preschoolers and participating in chores for children set the stage for the development of later skills and confidence. Facilitating both toddlers' and older children's decision-making ability is necessary for building skills needed for managing the challenges of SB [51]. Similar to school teachers assessing and addressing the unique learning needs of children with SB, clinicians and families should engage in intentional and targeted planning for self-management skill development. This plan needs to integrate and address the child's challenges with executive functioning, work-

ing memory, or other unique learning needs, as well as attend to the child's inherent strengths and interests.

The process of developing self-management skills is one that needs to be addressed early and include multiple supervised opportunities for practice in the home, healthcare provider's office, and broader community. Ideally, these building blocks are core components of clinical encounters and addressed in a systematic and incremental manner during both primary and specialty care visits or through other regular self-management interventions or programs specifically tailored for individuals with SB and their caregivers. It is also important to continue to work on developing these self-management skills incrementally as adolescents age into adulthood, while at the same time assessing and planning for any self-management supports a young adult with SB will need to maintain optimal health and independence as an adult.

Multiple research gaps were identified by the working group. A critical need is to understand what foundational skills and abilities in young children with SB facilitate the development of self-management and independence behaviors in later childhood and adulthood. Once those foundational skills are identified, interventions that optimize the development of these skills and abilities need to be evaluated. The complex timing and skills needed for incremental transfer of responsibility for condition self-management in adolescents is unknown. In addition, the multi-faceted and multi-level barriers and facilitators for developing autonomy, self-management, and independent living skills across the lifespan need to be better delineated. Increased attention should also be placed on testing interventions aimed at closing the gap between self-management behaviors in individuals with SB and their typically-developing peers. Similarly, family-centered interventions that identify the support needs of parents and facilitate parental transition from direct care to coach and

Table 3
Self-management instruments [84,85]

Name of instrument Citation	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			
KKIS-SB	– Twenty-two items.	Evidence reported [13]	– Parent scales are recommended.
Kennedy Krieger Independence Scales-Spina Bifida (KKIS-SB)	– Caregiver-reported measure of self-care skills for individuals ages 10 and above.	– Psychometric analysis with a sample of 122 parents of individuals with spina bifida ages 10–29.	– Child report version exists but psychometric data have not been published.
<i>The Kennedy Krieger Independence Scales-Spina Bifida Version: A Measure of Executive Components of Self-Management (KKIS-SB)</i> [13].	– Based on assumption that self-care skills require adequate executive functioning and that other scales do not assess the executive burden of these tasks.	Reliability	– Contact developers for more information on child-report KKIS-SB.
	– Four response pattern options:	– Internal reliability $\alpha = 0.89$	– 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).
	* More than 90% of the time	– Test-retest not reported	
	* 10–90% of the time	Validity	
	* Less than 10% of the time	– Exploratory factor analysis, reliability and construct validity using BRIEF (Behavior Report Inventory of Executive Function) were conducted.	
	* Not necessary or no opportunity	– Factor analysis supported two subscales.	
	– Two subscales: Initiation of Routines and Prospective Memory.	– Correlations between KISS-SB initiation of routines subscale and BRIEF summary scales ($r = -0.031-0.56$) 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.	
	– Initiation of Routines subscale items:	– Age-related changes and correlation with the Adaptive Behavior Assessment System scales also support validity of both KKIS-SB subscales.	
	* 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:		
	* Arrive at appointments on time		
	* Arrange transportation		
	* Self-examine for skin breakdown		
	* Start bowel program		
	* Perform pressure relief		
	* Write scheduled appointments		

Table 3, continued

Name of instrument Citation	Short description: Number of items; age range; type of instrument; subscales	Psychometric evidence: reliability and validity	Recommended use
AMIS II (Interview) <i>The Development of the Adolescent/ Young Adult Self-Management and Independence Scale-AMIS II: Psychometric Data</i> [46].	<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 * 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>Evidence reported [46]</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 $\alpha = 0.72–0.89$ - 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 young adults. Increase in self-management over one year was related to decrease in depressive symptoms. 	<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: 36-item Spina Bifida-specific instrument * AMIS II-SR-G: 28-item generic instrument

Table 3, continued

Name of instrument Citation	Short description: Number of items; age range; type of instrument; subscales	Psychometric evidence: reliability and validity	Recommended use
Medical Self-Management and Transition Readiness <i>Measurement of medical self-management and transition readiness among Canadian adolescents with special health care needs</i> [86].	<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 relating 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 [86]:</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 spina bifida) <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.

Table 3, continued

Name of instrument Citation	Short description: Number of items; age range; type of instrument; subscales	Psychometric evidence: reliability and validity	Recommended use
Spina Bifida Self-Management Profile (SBSMP) Sharing of Spina Bifida Responsibilities Scale (SOSBMR) Spina Bifida Independence Scales (SBIS) <i>Condition self-management in pediatric spina bifida: A longitudinal investigation of medical adherence, responsibility-sharing, and independence skills</i> [23].	<ul style="list-style-type: none"> - A series of 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 (yes, no, not sure, or NA) but does not measure if the child does them on a consistent basis. 	<p>Evidence reported [23]:</p> <ul style="list-style-type: none"> - These measures reported in a study of 140 children with spina bifida 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 > 0.60) 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. - 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. 	<ul style="list-style-type: none"> - May have promise for future use, especially for comparison to diabetes. - Several scales are needed to measure these concepts and may have item burden in clinical practice.
Generic Self-Management/Transition Readiness Instruments. No reported use in spina bifida TRAQ (Transition Readiness Assessment Questionnaire) <i>Measures the transition-readiness of youth with special health care needs</i> [69].	<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 reported [69]</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.

Table 3, continued

Name of instrument Citation	Short description: Number of items; age range; type of instrument; subscales	Psychometric evidence: reliability and validity	Recommended use
<p>UNC TR(x)ANSITION scale. <i>A clinical tool to measure health care transition components from pediatric to adult care</i> [87].</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 [87]:</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> [60,61].</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 [60]:</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 < 0.0.99, 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 [88]. - 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.

Table 3, continued

Name of instrument Citation	Short description: Number of items; age range; type of instrument; subscales	Psychometric evidence: reliability and validity	Recommended use
STARx Self-Management and Transition to Adulthood with Rx = Treatment <i>Self-management and transition readiness assessment: development, reliability, and factor structure of the STARx questionnaire</i> [89].	<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. * Subscales: <ul style="list-style-type: none"> * Medical Management * Provider Communication * Disease Knowledge * Adult Health Responsibilities * Resource Utilization 	<p>Evidence Reported [89,90]:</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 ($\alpha = 0.44$ 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 0.31 to 0.88. - 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. 	<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.

Legend: Table 3 used with permission and obtained from Guidelines for the Care of People with Spina Bifida 2018: (<https://www.spinabifidaassociation.org/guidelines/>). Additional resource: 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: <https://www.spinabifidaassociation.org/resource/beyond-crayons-resource-packet/>.

consultant are presently limited in scope and availability. It is also important to determine if routine clinical assessments of self-management behaviors that result in the development of action plans tailored toward the adolescent and their family yield improved outcomes. It is unclear if these types of interventions enhancing self-management and independence can be delivered in the clinical setting or if alternative structures need to be developed, such as implementation within school settings. Further, the optimal structures for coordinated, comprehensive transition to adult care have yet to be determined. As longevity in SB increases, understanding the self-management and independence needs in young, middle-age, and older adults with SB as they age becomes imperative. Finally, the role of the health care provider and the larger health care system in optimizing self-management and independence outcomes needs to be explored.

Addressing these gaps should provide a map for future care-providers to effectively help families prepare children with SB to become independent. In the meantime, it is hoped that these guidelines will steer caregivers to have developmentally appropriate expectations for the development of self-management skills. 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 [52–54]. These findings will need to be integrated into ongoing care.

Two additional publications build on Self-Management and Independence Work Group's work. Both reflect the importance of assessment in order to meet the person with SB at the appropriate readiness level for change in this area [55,56].

Acknowledgments

This edition of the *Journal of Pediatric Rehabilitation Medicine* includes manuscripts based on the most recent "Guidelines For the Care of People with Spina Bifida," developed by the Spina Bifida Association. Thank you to the Spina Bifida Association for allowing the guidelines to be published in this forum and making them Open Access.

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

The authors declare no conflicts of interest.

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