

Guidelines for the Care of People with Spina Bifida

13-17 years 11 months

Care Coordination Guidelines

1. It is recommended that the Spina Bifida Care Coordinator provide developmentally-appropriate care education across the spectrum of symptoms and conditions related to Spina Bifida to better empower children and families to manage their own care and recognize complications and emergencies. Identify and or improve gaps in the family knowledge base specifically related to the teenage age period (mobility progress, skin inspection, bowel and bladder care, sexuality, academic/cognitive development, social functioning at school and with peers, high risk behaviors, and more). (clinical consensus) (Bowel Function and Care Guidelines, Mental Health Guidelines, Mobility Guidelines, Neuropsychology Guidelines, Skin (Integument) Guidelines, Urology Guidelines)
2. It is recommended that the Spina Bifida Care Coordinator monitor secondary school functioning and update the school education and health plan so that it includes preparation for college or other higher education opportunities. Encourage participation in age-appropriate activities with peers outside of school. Where appropriate, provide information for driver education and training programs for the teenager with Spina Bifida. (clinical consensus)
3. It is recommended that the Spina Bifida Care Coordinator communicate with the family and multidisciplinary Spina Bifida care team to ensure the individual with Spina Bifida is up-to-date on all sub-specialty care visits, imaging, monitoring, and equipment needs where appropriate. This may include assistance with insurance authorization when needed.
4. It is recommended that the Spina Bifida Care Coordinator update the primary care provider on the current care goals and recommendations of the Spina Bifida multidisciplinary care team. The coordinator should use two-way communications to identify and address medical concerns and obtain updated records from the primary care provider such as immunizations, growth charts, developmental screenings, and other materials.
5. It is recommended that the Spina Bifida Care Coordinator work with the teenager and his/her families, Spina Bifida team members, and therapists to continue progress on self-management goals and education. Monitor family progress at regular intervals in self-management and clinic visits with the goal of achieving as much independence as is realistically possible. Engage the school nurse to help facilitate self-management and independence. 14 Teach self-advocacy and encourage the teenager to participate as much as possible in his or her own self-management. When appropriate, discuss what limitations to independence the teenager may have due to deficits in memory, cognition, and executive functioning and provide the parents with additional resources and support services, as needed. 17-19 (Self-Management and Independence Guidelines)
6. It is recommended that the Spina Bifida Care Coordinator begin preparing the teenager for transition to adult health care, including familiarizing them and their family with the Transition Guidelines and Self-Management and Independence Guidelines. Encourage the family to develop and assemble their own health care folder and records for use during travel,

appointments in hospitals that are away from their home area, and other occasions when they will be away from the medical home. Encourage the person to make his or her own medical appointments once she or he is capable of doing so, and to start leading the conversation with specialists and other providers during clinic visits. Assist the family by making them aware that transition to adult life involves many aspects beyond health care, including educational planning or job training, making arrangements to live independently, and financial planning.

(Self-Management and Independence Guidelines, Transition Guidelines)

7. It is recommended that the Spina Bifida Care Coordinator assess family dynamics in coping with the diagnosis and evaluate their psychosocial stressors. Collaborate with primary care provider to review age-appropriate screenings and assist with referrals to mental health and social services when appropriate. (clinical consensus) (Mental Health Guidelines)

8. It is recommended that the Spina Bifida Care Coordinator serve the family as the lead contact person and information provider for the multidisciplinary medical services for the person with Spina Bifida and monitor family needs and prescriptions for durable medical equipment, supplies, and medications, as needed. (clinical consensus)

9. It is recommended that the Spina Bifida Care Coordinator conduct an inventory of the person's ability to provide self-management, complete activities of daily living, and manage mobility equipment and transportation needs. For a person with a significant intellectual disability who may not be able to live independently, assist the family with the conservatorship process prior to age 18, and with maintaining Supplemental Security Insurance (SSI) and other types of insurance coverage. (clinical consensus) (Self-Management and Independence Guidelines)

Health Promotion and Preventive Health Care Services Guidelines

1. Monitor that the child is making routine well-child visits to his or her primary care provider to receive age-appropriate health promotion and preventive services, including age-appropriate screenings for:

- o Hypertension. Since there is no agreement on blood pressure targets for patients with Spina Bifida, it is recommended that baseline blood pressure is monitored to know what is considered hypertensive for the particular child. (clinical consensus)
- o Iron deficiency.

- o Lipid disorders.

- o Overweight/obesity, including the role in hypertension. (Nutrition, Metabolic Syndrome, and Obesity Guidelines)

- o Abuse, neglect, and/or violence. (Family Functioning Guidelines)
- o Social Isolation, Anxiety, Depression. (Mental Health Guidelines)

- o Motor vehicle and wheelchair safety. (Mobility Guidelines)

- o Contraceptive use, pregnancy, and sexually transmitted diseases. (as age appropriate) 1 (Sexual Health and Education Guidelines, Women's Health Guidelines, Men's Healthcare Guidelines)

2. Provide counseling about tobacco and illicit drug use and refer the family to an appropriate treatment program if needed.

3. Provide guidance on skin cancer prevention.

4. Provide information about adaptive physical and recreational activities keeping in mind the particular child's degree of mobility.⁷ (Physical Activity Guidelines)

5. Provide information about accessible physical activity and recreational opportunities in the community.⁹ (Physical Activity Guidelines)

6. Monitor for pain and changes in pain using an appropriate pain scale for the child's level of cognition and communication, as pain may not be clearly recognized due to the unique neurologic status of children with Spina Bifida. Be aware that depending on their cognitive status, the child may not be able to give specific answers to questions such as the severity, frequency and duration of the pain. Proceed with appropriate evaluation and treatment.

7. Monitor for comorbid conditions that are specific to children with Spina Bifida, both during visits that are specifically intended to monitor Spina Bifida conditions as well during well-child visits. (Neurosurgery Guidelines, Orthopedics Guidelines, Bowel Function and Care Guidelines, Urology Guidelines, Mobility Guidelines, Skin Care Guidelines)

- o Shunt concerns. Ask about any neurologic changes.

- o Sleep apnea. Ask if sleeping is restful and if there are snoring or apneic pauses during sleep.

- o Skeletal and limb deformity. Check for new issues with bracing, positioning, or function. (clinical consensus)

- o Constipation, urinary tract infections (UTIs), renal function, and problems with bowel and bladder regimens. Provide prescriptions for routine bowel medications, treatment of recurring UTIs, monitor for adherence to bowel and bladder management program and changes in bowel/bladder function. (Bowel Function and Care Guidelines, Urology Guidelines)

- o Skin breakdown and pressure injury. Urge the family and child (if appropriate) to perform daily skin checks. Recommend that the child's skin is properly moisturized, and that appropriate weight-shifting is taking place based on the child's neurologic level. (Mobility Guidelines, Skin Care Guidelines)

- o Adaptive equipment needs, including for orthoses, crutches, walkers, and wheelchairs. Make referrals to necessary subspecialists. (clinical consensus) (Mobility Guidelines)

- o Osteoporosis. Encourage weight-bearing activities for at least one hour per day to promote bone health as well as for its social benefits. (clinical consensus) (Mobility Guidelines, Orthopedics Guidelines, Physical Activity Guidelines)

8. Promote care coordination between Spina Bifida-specific subspecialists and primary care providers. (Care Coordination Guidelines)

9. Educate families on early signs of chronic conditions related to Spina Bifida.

Transition Guidelines

1. Provide updates for children and families regarding a probable trajectory for adult function and expectations for optimal independence according to the individual's abilities and chronic condition status.

2. Consider neurocognitive assessment to identify cognitive, adaptive, or learning support needs if knowledge or skill gaps are identified. (Neuropsychology Guidelines, Self-Management and Independence Guidelines)

3. Discuss transition planning with children and families including:
 - a. Expectations of when the transfer to adult care will occur based on the individual's health condition, insurance/funding, cognitive development, and personal/family needs.
 - b. Expectations of who will provide care throughout the transition process.
 - c. Counselling regarding long-term financial, insurance, and supportive living (housing and transportation) plans, based on the individual's current needs and probable trajectory of adult function.
 - d. Information regarding the child's education and employment needs, such as vocational rehabilitation services, school transition planning as part of the Individualized Educational Plan [INSERT LINK], and adaptive vocational needs.
 - e. Preparation for decision-making supports and modalities that maximize the individual's ability to participate in decisions for themselves once they are age 18, such as a medical power of attorney, supportive decision-making, or guardianship. Referral for neurocognitive testing and to medical legal partnerships may be needed.
 - f. Information regarding the Social Security Administration's Disability Determination Services before age 18, as applicable.
 - g. Creation of a medical summary including past medical and surgical history, current care plans, medications, allergies, vaccines, and current providers.
 - h. Self-management support. Consider using transition and self-management assessment tools to direct goals and interventions. (Self-Management and Independence Guidelines)
4. Ensure that the patient's views and preferences are included in transition planning.
5. Designate time alone with the child for at least part of their visit, if developmentally appropriate.
6. Consider having a designated transition clinic or care coordinator to support transition planning and coordination.
7. Ensure patient-centered and developmentally appropriate preventive and chronic condition management services are provided throughout transition. (Health Promotion Guidelines) Evaluate management plans and assess for necessary adaptive equipment and supplies to maximize independent function.

Family Functioning Guidelines

1. Provide support and ongoing counseling for parents, child, and siblings, as needed.
2. Be aware that although interventions should target all families, some families are particularly at-risk for adjustment and adherence difficulties (e.g., those from lower socioeconomic backgrounds and single-parent families).
3. Assess parent-child communication and their relationship. Aid parents to encourage the development of autonomy in their child with Spina Bifida. (Self-Management and Independence Guidelines)
4. Encourage the family to begin planning for their child's transition to adult health care. (Transition Guidelines)

5. Begin discussions of other important developmental milestones, including educational and vocational achievement, living independently, and community participation. (Transition Guidelines)
6. Give advice to the child and family about the right to free and appropriate education in the least restrictive environment through the public schools (i.e., explain services available under the Individuals with Disabilities Education Act and Section 504 of Vocational Rehabilitation Act of 1973). (clinical consensus)
7. Assist with normative sexual education, as well as specific issues relevant to the teen's condition. Work with the teen to navigate sexual expression in a safe and mature fashion. (Sexual Health and Education Guidelines)
8. Continue to encourage the family to facilitate medical self-management in their child with Spina Bifida. (Self-Management and Independence Guidelines)
9. Assess the family context for helping the child to develop self-management skills and to carry out medical regimens and identify possible barriers to adherence. (Self-Management and Independence Guidelines, Transition Guidelines)

Mental Health Guidelines

1. Assess peer relationships and friendships.
2. Assess for at-risk behaviors (alcohol, drug, or tobacco use and unsafe or unprotected sex), and identify areas of strength and build on resources that encourage resilience.
3. Screen for depression or anxiety and initiate individual and family interventions when appropriate.
4. Provide counseling and/or behavioral support as needed. (clinical consensus)
5. Promote transfer of medical responsibility from parent to child in those who have the requisite abilities and cognitive capacity. (Self-Management and Independence Guidelines)
6. Refer for social skills training as needed.
7. Encourage activities and hobbies that improve face-to-face social contact. (clinical consensus)
8. Provide counseling regarding sexuality, sexual functioning, fertility, and contraception. Focus on sexual safety issues. (Sexual Health and Education Guidelines)
9. Discuss the safe use of and choices around drugs and alcohol and conduct risk assessment in this domain.
10. Discuss the importance of initiating and organizing opportunities for social activities.
11. Discuss the relationship between independence and interdependence and mental health. (clinical consensus) (Self-Management and Independence Guidelines)
12. Facilitate the child's involvement with a peer role model, such as a teen with Spina Bifida who is of a similar age). (clinical consensus)
13. Provide or refer to opportunities for formal or informal mentoring. (clinical consensus)
14. Encourage the teen to participate in the school's IEP transition team meeting. (clinical consensus)
15. Develop a plan for the teen's transition to independent living, post-secondary education, vocational training, and career interests. (clinical consensus) (Transition Guidelines)
16. Develop a plan for transition from pediatric to adult health care. (clinical consensus) (Transition Guidelines)

Quality of Life Guidelines

Psychosocial well-being

1. Assist families in their efforts to facilitate the development of protective beliefs (e.g. hope, optimism, attitudes, future expectations, active coping strategies) and behaviors such as showing affection, bouncing back when things don't go their way, showing interest in learning new things, handling negative situations, and establishing and maintaining friendships. (Mental Health Guidelines, especially the section on peer relationships)
2. Consider strategies to assess and strengthen family functioning, which can be of critical importance in QOL outcomes in children. (Family Functioning Guidelines)
3. Consider strategies to optimize peer relationships. (Mental Health Guidelines)
4. Consider each individual's unique priorities important in QOL. (clinical consensus)
5. Refer to community resources such as sports, camps, scouts, and other community programs that enhance protective factors. (clinical consensus) (Self-Management and Independence Guidelines)
6. Address strategies to compensate for executive functioning challenges. (Neuropsychology Guidelines)

Continence/mobility

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

Pain

1. Evaluate presence and characteristics of any pain experienced.
2. Develop strategies to address pain and its impact on school, work, recreation, and social activities. (clinical consensus)

Measurement

1. Use a systematic approach to evaluating QOL/HRQOL.
2. Consider using both self and parent-report instruments.
3. Use the new Spina Bifida HRQOL instrument that measures perception ("concerned about," "worried about") and avoids the problem of focusing on function in the physical domain (walking long distances, climbing stairs, jumping) when assessing children with Spina Bifida. Omit any measure that captures the impact in the physical domain. Emotional, social, and school/cognitive domains in most perception-based instruments are useful. (Appendix 1)
4. Use an age- and condition-specific instrument to assess QOL/HRQOL. (Appendix 1)
5. Evaluate both the child's self-report and the parent report of QOL/HRQOL. If assessment time is limited choose self-report.
6. Consider using a single-item QOL question(s) with follow up assessment if needed.(Appendix 1). For example:
 - "How would you rate your quality of life?"

- “What makes up QOL for you?”
- “What do you think would make your QOL better?”

Self Management and Independence 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-efficacy 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.
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.
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.
 - Encourage increasing responsibility for behaviors such as management of medication, bowel and bladder programs, and skin-monitoring. (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).
 - Evaluate and monitor cognitive functions as they underpin decision-making, goal-setting, self-regulation, self-management, socialization, and transition issues. (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. (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.
 - Encourage the family to expand the range of responsibilities for daily life activities, chores, and jobs.
 - o 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). (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).
10. Encourage the use of technology to enhance self-management.
11. Share expectations and resources for future independent living, transition to college or employment.
12. Provide consultation to adult providers with limited skill in providing care to those with congenital conditions such as Spina Bifida. (Transition Guidelines)

Neuropsychology Guidelines

1. Promote interventions that address integration and assimilation of information with a specific focus on reading comprehension and mathematics problem-solving, as well as other areas of applied mathematics and functional numeracy. Intervention programs should be maintained because the absence of intervention is associated with plateaus in skill development in most populations with disabilities.
2. Encourage participation in school-related and extracurricular activities and create vocational plans and transitional services with a focus on functional independence. (clinical consensus)
3. For students receiving special education services, the Individualized Education Plan [INSERT EARLY INTERVENTION LINK] (IEP) is required to include a formal transition plan to address vocational, occupational, and life skill domains by 14 to 16 years of age. Coach parents to ask about educational transition plans and to request evaluations to bolster the plans. Early transition plans are essential to develop the capacity to assume the roles and responsibilities of the post high school environment and achieve optimal independence. They are also needed to ensure that appropriate referrals are made to adult agencies, that there is suitable life and vocational skill training, and that there are discussions about plans after high school. Educate families on the need for a transition plan and check to ensure a comprehensive plan is created. If needed, refer to state-based educational advocacy programs (e.g., the ARC) that can provide support and education. (Transition Guidelines)
4. Because social skills of individuals with SBM are strongly related to neuropsychological variables, namely attention and executive function, consider using psycho-educational and/or neuropsychological assessments to inform psychosocial interventions and mental health supports.³⁹ (Mental Health Guidelines)
5. Be aware that in addition to the cognitive and learning problems associated with the underlying neurological disorder, persons with SBM may experience reduced quantity and quality of social interactions. Encourage structured opportunities for social interaction through

school, church, and after school opportunities. (clinical consensus) Conduct yearly screening and timely referrals for appropriate diagnosis and treatment of anxiety and/or depression with psychotherapy and/or medication treatment as needed. (Mental Health Guidelines, Quality of Life Guidelines)

6. Identify cognitive strengths and weaknesses for those who are assuming responsibility for their own medical care. This may require formal assessment, particularly if children are unable to assume responsibility for their own medical decision making and will require guardianship. Efforts to assess and build communication skills, increase knowledge about their medical condition and history, and develop medical triaging skills needs to begin as early as possible because it may take children in this age group over several years to learn the skills necessary to understand and take responsibility for their own medical care. Address bladder and bowel incontinence, as both can be major issues affecting social adjustment. (clinical consensus) (Bowel Function and Care Guidelines, Transition Guidelines, Urology Guidelines)

7. Advise children and/or their parents/guardians to obtain copies of psycho-educational and/or neuropsychological assessments. Explain that documentation of intellectual disability and/or learning disability prior to age 18 is needed to qualify for services in adulthood. A diagnosis of intellectual disability requires thorough assessment of adaptive skills. This is an important point because school programs and special education service evaluations may not always include formal assessment of adaptive skills.

Neurosurgery Guidelines

Patient/Family

1. Observe the child for clinical signs of shunt failure, brainstem dysfunction, TSC, and/or syringomyelia. (clinical consensus)
2. Continue to foster a working relationship with the team of Spina Bifida providers. (clinical consensus)
3. Neurosurgery should assist child and family in learning the concept of transition to adult care and in identifying an adult neurosurgery provider. (clinical consensus) (Transition Guidelines)

Providers/Neurosurgeons/Spina Bifida Clinic

1. Follow children ages 13-17 years 11 months at 12-month intervals in a Spina Bifida clinic. (clinical consensus)
2. Begin to address transition to adult neurosurgical provider early in teen years to promote self-knowledge and functional independence and encourage teen self-monitoring. (See Transition and Self-Management and Independence Guidelines)
3. Review and observe for signs of acute shunt failure (headache, neck pain, vomiting, lethargy/sleepiness), and chronic shunt failure (recurring low grade headache and neck pain, behavioral and/or cognitive changes, neurological decline, urological changes, and increasing orthopedic deformities and/or progressive scoliosis). Follow the child clinically to observe for these signs.
4. Review with the family and child the signs of brain stem dysfunction that might occur in this age range (poor control of secretions, swallowing dysfunction, stridor, and declining language function). Follow the child clinically to observe for these signs. (clinical consensus)

5. Teach or review with the family and child and urge them to observe for signs of TSC (back pain, declining sensorimotor function, urological changes, and progressive orthopedic deformities and/or scoliosis). Follow the child clinically to observe for these signs.
6. Teach or review with the family and child and urge them to observe for signs of syringomyelia (back pain and sensorimotor changes in arms and hands). Follow the child clinically to observing for these signs. (clinical consensus)
7. Use adjunctive studies judiciously (imaging such as MRI/CT, urodynamics, and sleep and swallow studies) during routine visits with the well child, according to experience, preference and best clinical judgment, to augment clinical decision-making. (clinical consensus)

Mobility Guidelines

1. Assess neurologic level and strength changes using standardized assessment tools at each clinic visit. Monitor for changes in gait, sensation, bowel and bladder function, and musculoskeletal changes. (clinical consensus)
2. Monitor ambulation or wheelchair mobility. If ambulation is declining, suggest alternate mobility options. (clinical consensus)
3. Continue therapy or home programs to maintain mobility goals, emphasizing flexibility, range of motion, and overall strengthening. (clinical consensus)
4. Verify that the teenager knows how to check insensate skin, especially after activity, and how to ameliorate friction and pressure. (clinical consensus) (Integument (Skin) Guidelines)
5. Optimize gait with supportive orthoses or devices for balance. Monitor for torque forces on the joints or excessive forces in the upper body.
6. Explore the best mobility option with the teenager and have a frank discussion about the risks and benefits of all systems. (clinical consensus)
7. Monitor for a secondary injury and, if identified implement a prevention program. Areas at risk of secondary injuries for children who walk are the knees and ankles and the shoulders and wrists in those who use a wheelchair. (Orthopedic Guidelines)
8. Recommend therapy interventions to maintain mobility, if there is a change in functional status. (clinical consensus)
9. Collaborate with orthopedic specialists to monitor for age specific musculoskeletal problems. (Orthopedic Guidelines)

Orthopedics Guidelines

1. Monitor for the development or progression of scoliosis clinically, with radiographs as necessary, if indicated by the physical exam. Perform radiographs in a sitting in position in those who can sit but not stand and in a standing if position in those who can stand. If the curve has progressed to an operative magnitude (50 degrees), discuss the risks and benefits of surgical treatment with the family. (clinical consensus)

2. Monitor for deterioration of gait and consider treatment of orthopedic deformities leading to deterioration such as hip and knee contracture or rotational deformities. Computerized gait analysis may be useful for decision-making in the case of children with low lumbar and sacral level lesions.
3. Conduct a history and physical examination (with radiographs, if indicated) on an annual basis, unless greater frequency is indicated. (clinical consensus)

Physical Activity

1. Discuss with children the benefits of participating in physical activity, recreation, and sports. Discuss with children and parents/caregivers the importance of limiting sedentary behaviors. Encourage children and parents/caregivers to consider choices about where they can be actively engaged with peers who have and those who don't have a disability. (Health Promotion and Preventive Health Care Services Guidelines)
2. Recommend that children and parents/caregivers follow the National Physical Activity Guidelines as closely as possible, unless a health care provider advises that they are medically unsafe. (clinical consensus)
 - Engage in 60 minutes of physical activity or more each day.¹⁵
 - Aerobic activity should make up most of the child's activity each day; vigorous intensity aerobic activity should be done at least 3 days/week.
 - Muscle strengthening activities should be done at least 3 days/week as part of the 60 or more minutes.
 - Bone strengthening activities should be done at least 3 days/week as part of the 60 or more minutes
3. Use health care encounters and follow-up meetings to develop physical activity goals and monitor progress (e.g. minutes of physical activity per day). Use motivational interviewing techniques with children and parents/caregivers to talk about physical activity goals and work through barriers. Support children and parents/caregivers to develop an action plan with strategies to support the participation of children age 13-17 years in physical activity in their community and school. Prescribe, using a prescription pad, physical activity based on goals identified by children with Spina Bifida. (clinical consensus)
4. Perform pre-participation evaluations for children with Spina Bifida in collaboration with parent/caregivers, pediatric specialists, therapists, coaches, and others to identify medical risks and modifications that can be made to ensure participation.
5. Identify strategies to minimize risks of illness and injury related to participation through activity adaptations and safety precautions. Identify and provide additional support and information for children age and parents/caregivers on precautions to take when children with shunts and ambulatory limitations are being active. Discuss strategies that balance the parents'/caregivers' involvement with their child's need for independence when they participate in physical activity.
5. Use a team approach and include PTs/OTs to work with parents/caregivers to ensure their child has proper fitting mobility equipment to maximize physical activity participation. (clinical consensus)

6. Work with children with Spina Bifida and their family to address personal barriers such as bowel/bladder care, medical events, assistive devices, as well as environmental factors that may affect participation.
7. Advocate for and address barriers to participation of children with Spina Bifida in physical activity, recreation, and sports.
8. Inform parents/caregivers of their child's rights to adapted physical education/activity and encourage children and parents/caregivers to advocate for physical activity goals to be added to their IEP or Section 504 plan (if eligible for IEP or Section 504 plan).
9. Assist students who are considering post-secondary education to assess supports for physical activity in the educational institutions they are considering and include these in their individual transition plans.
10. Advocate for the participation of children with Spina Bifida in both unified and adapted sports, recreation, and physical activity programs.
11. Provide families with a local/regional therapeutic recreation and adapted sport resource guide.
12. Discuss with children where they feel most comfortable being physically active and about their options (e.g., in the community, at school, with peers with/without disabilities).
13. Highlight that ages 13-17 years is a critical period to build physical activity into a daily routine that will preserve overall lifelong satisfaction and community participation among persons with Spina Bifida.

Men's Health Guidelines

1. Conduct an annual scrotal exam that documents testicular position, size, consistency, symmetry, and presence or absence of masses. (clinical consensus)
2. Access and document genital sensation (penile, scrotal) and Tanner staging annually.
3. Instruct men about monthly testicular self-examinations (TSE). (clinical consensus)
4. Initiate open-ended conversations with boys age 13-17 with Spina Bifida about their knowledge of normal sexual function when the provider deems that the boy is developmentally ready, or when there is evidence of sexual curiosity and experimentation in their medical history.9-12 (clinical consensus) (Sexual Health and Education Guidelines)
5. Educate patients that sexual function may be altered as a sequela of Spina Bifida.(clinical consensus)
6. Provide information about safe sexual practices and genetic risk factors. (clinical consensus) (Sexual Health and Education Guidelines)
7. Offer human papillomavirus (HPV) vaccination per Centers for Disease Control and Prevention and American Academy of Pediatrics guidelines, as appropriate.
8. Characterize and document erectile function when it becomes developmentally appropriate. Providers may use the International Index of Erectile Function (IIEF) or Sexual Health Inventory for Men (SHIM). (clinical consensus)
9. When relevant, characterize and record orgasmic and ejaculatory function. (clinical consensus)

Sexual Health and Education Guidelines

1. Acknowledge that sexual health is an important part of life. (clinical consensus)
2. Discuss healthy relationships in gender-neutral language as the teen years are the time when many achieve self-awareness about sexual orientation.
3. Educate teens about intimate partner violence and sexual assault.
4. Discuss safe-sex practices including the use of non-latex condoms to prevent sexually transmitted infections and unwanted pregnancies.
5. Refer to a women's health provider such as a gynecologist, adolescent medicine specialist, or family medicine practitioner if the teen with Spina Bifida intends to become sexually active. Refer young men to a sexual function clinic if desired. (clinical consensus) (Men's Health Guidelines, Women's Health Guidelines)
6. Ensure that the Guidelines for Adolescent Preventive Services are implemented.
7. Create an environment in which the teen feels comfortable and safe discussing sexual health, including being able to speak to them alone and confidentially.
8. Educate parents by presenting them with factual information and encourage them to provide developmentally appropriate sexual education to their children.
9. Encourage parents to discuss information that their children are receiving about healthy relationships from school, their peers, the media, and social media.
10. Discuss sexuality routinely and openly during health care visits, and acknowledge the fluidity of sexuality and gender.
11. Allow the teen to ask questions about sexual development and sexuality.
12. Serve as a resource to schools to ensure that children with Spina Bifida participate in sexual education.
13. Underscore the goal of continence (Bowel Function and Care Guidelines, Urology Guidelines) for optimal sexual relationships in the future. (clinical consensus)
14. Provide education about pubertal development and evaluate pubertal development and any abnormal physical findings (Endocrine: Puberty and Precocious Puberty Guidelines).
15. Educate teens and parents regarding birth control options, pregnancy, genetic risk, and sexually transmitted infection risk associated with sexual activity including the use of non-latex barrier methods. (Men's Health Guidelines, Women's Health Guidelines)

Urology Guidelines

1. Obtain a renal/bladder ultrasound yearly, if the child is stable. (clinical consensus)
2. Obtain a renal/bladder ultrasound as needed, if the child has recurring symptomatic UTIs or if urodynamic testing identifies bladder hostility. (clinical consensus)
3. Obtain a serum creatinine test yearly. If the child has low muscle mass, consider an alternative measure of renal function.⁵ (clinical consensus)
4. Obtain serum chemistries including B12 yearly on any child who has had urinary reconstruction. (clinical consensus)
5. Transition urologic care to self-management, if doing so is developmentally appropriate for the child.¹⁴⁻¹⁵ (clinical consensus) (Self-Management and Independence Guidelines)

6. Transition bowel program to self-management, if doing so is developmentally appropriate for the child. (clinical consensus) (Bowel Function and Care Guidelines)

Women's Health Guidelines

1. Manage pelvic organ prolapse, which can occur at any stage of life in women with Spina Bifida, in consultation with an urogynecologist. Take into account the possibility of decreased pelvic sensation.
2. Provide guidance on reproduction, sexual health and education. (Sexual Health and Education Guidelines)
3. Contraception options should be made available and discussed to in a non-judgmental manner, taking into account health concerns such as decreased mobility, risk of decreased bone mineral density, latex allergy and use of antiepileptic medications and genetic risk factors. (clinical consensus) Consider consulting a gynecologist in a complex scenario.
4. Offer HPV vaccination per Centers for Disease Control and Prevention and American Academy of Pediatrics guidelines, if appropriate.

Bowel Function and Care Guidelines

1. Discuss consequences of constipation and bowel incontinence (including shunt malfunction, urinary incontinence, UTIs, skin breakdown, social isolation) and focus on developing independent management skills. (Self-Management and Independence Guidelines)
2. Establish or maintain the goal of bowel continence and institute or maintain the bowel continence program using the guidelines below.
3. Assist the child with learning how to minimize and manage bowel accidents.
4. Use barrier creams to protect perineal area from breakdown as needed.
5. Keep a bowel habit diary to better understand triggers for incontinence and overall patterning to direct a choice of options for bowel management.
6. Focus on fiber, fluids, exercise, and timed bowel movements after meals.
7. Consider a twofold attack of oral and rectal interventions to meet the goal of bowel continence without constipation.
8. Use dietary management (fiber, fiber supplements, and fluids), pharmacologic adjuncts (sennoside, polyethylene glycol), and/or rectal stimulants (glycerin, docusate sodium, or bisacodyl suppositories) to manage constipation.
9. Discuss other options for treatment if the above have failed, including cone enema or other transanal irrigation, cecostomy, or antegrade continence enema (Malone).
10. Refer to Spina Bifida clinic or specialist with expertise in bowel management in Spina Bifida. (clinical consensus)
11. Access support services for personal care, if needed.

Endocrine: Puberty and Precocious Puberty Guidelines

1. Monitor and document weight and height velocity closely at every health supervision visit. Height should be measured (if possible) using a stadiometer. Often there may be difficulty assessing height due to inability to stand, scoliosis or contractures. In these cases, arm span or

another appropriate parameter may be used. Care should be taken to use the same parameter at subsequent visits. (clinical consensus)

2. Perform a complete physical exam, including of the breasts and genitalia, at each health supervision visit. Offer for the exam to be completed by a provider of the same sex if the child and/or family is more comfortable with a same-sex provider. (clinical consensus)
3. Document all positive and negative findings of the physical exam. (clinical consensus)
4. Discuss the outcomes of the evaluation with the parents or caregivers and ask them if they have any concerns. (clinical consensus)
5. Refer the child to a pediatric endocrinologist if there is clear evidence of abnormal timing, tempo, or sequence of pubertal development. (clinical consensus)
6. Consider a referral to a mental health professional if the child is having psychosocial issues with his or her growth or development. (clinical consensus)

Integument (Skin) Guidelines

1. Inspect skin daily. Explore the teen perceptions of self-efficacy for skin checks and barriers to skin checks. Develop plans to increase self-efficacy, if needed.
2. Suggest children and adults who use wheelchairs to use a pressure-relieving cushion and check it daily.
3. Identify and discuss risk factors that specifically increase the risk of pressure injuries in children and adults with Spina Bifida, such as using a wheelchair, having had surgery above the knee, shunts, a higher level of lesion, recent surgery, bladder incontinence, and being of the male gender.
 4. Review with the caregiver, child, or adult the consequences of heat, moisture, or pressure related to insensate areas.
5. Teach parents/caregivers/child/adult how to inspect for well-fitting orthoses.
6. Discuss the need to check water temperature and encourage the use of a bath water thermometer.
7. Tell children/adults to check for hot surfaces that have been exposed to the sun such as car seats.
8. Promote adequate hydration and proper nutrition for healthy skin. (Nutrition, Metabolic Syndrome, and Obesity Guidelines)
9. Encourage parents, caregivers, children, and adults to keep skin clean and dry.
10. Suggest wearing seamless socks that are clean and dry.
11. Suggest the use of antiperspirant on areas with perspiration, such as the feet and intertriginous areas.
12. Encourage seeking treatment if the skin is compromised.
13. Advise children and adults who are non-ambulatory and use a wheelchair to engage in pressure-relieving activities every 15 minutes.
14. Teach safe transfer skills to non-ambulatory patients.
15. Seek treatment immediately for any pressure injury. Refer to wound clinic for any pressure injury at stage three or greater.

Latex and Latex Allergy Guidelines

1. Educate teens directly about avoidance of latex products including latex-containing urinary catheters and educate them to know about safe latex-free alternatives. (clinical consensus) (Appendix 1)
2. Discuss avoidance of rubber balloons at parties, school activities, restaurants, and other gathering places for events.
3. Teens identified as having a latex allergy should have diphenhydramine and self-administered epinephrine available at all times. (clinical consensus)
4. Instruct families to check food preparation in public venues as it should be prepared with latex-free gloves. (clinical consensus)
5. Educate teens about latex-safe contraceptive products before they decide to become sexually active. (clinical consensus) (Sexual Health and Education Guidelines) (Appendix 1)
6. Urge children to continue following latex precautions because risk-taking during the teen years is common. (clinical consensus)
7. Review principles of latex precaution with the teen during a clinic visit and answer any questions. (clinical consensus)
8. If a latex allergic patient does not know if he or she is allergic to cross-reacting foods and has had anaphylaxis to latex exposure, it may be prudent for an allergist to test the patient. If a positive test is found, then a food challenge would be indicated in the case where there is no history of food related clinical reaction. (Appendix 2) Many of the positive tests may be due to laboratory cross-reactivity, but a clinical response of allergy will not be provoked.

Nutrition, Metabolic Syndrome and Obesity Guidelines

1. Conduct annual assessments of weight, height or arm span, and BMI. (clinical consensus) (Appendix: BMI and Body Composition Measurements)
2. Consider monitoring other measures of adiposity, such as waist circumference. (Appendix: BMI)
3. Conduct annual assessment of blood pressure/percentiles to monitor for pre-hypertension and hypertension. (clinical consensus)
4. Provide opportunities for teens and parents to talk about their priorities and concerns regarding nutrition and weight.
 - Discuss how nutrition can play an important role in helping individuals with Spina Bifida maintain a healthy weight; minimize skin breakdown, and increase activity and endurance.
 - Discuss that children with Spina Bifida, especially those who are non-ambulatory, who undertake low levels of physical activity, and who have higher body fat levels or contractures, are at increased risk for bone fractures. Recommend a diet with adequate calcium and vitamin D.
 - Provide regular opportunities for teens to discuss any concerns with their weight, growth and diet. A trusting therapeutic relationship can greatly facilitate an honest and open discussion.
 - Identify the teen's priorities and negotiate goals that meet those priorities as well as the parent's and clinician's goals
 - Use a strengths-based approach that highlights their nutritional achievements and celebrates successes.

- Discuss with parents, if relevant, that the Body Mass Index (BMI) is an imperfect indicator of health in all young people and especially in children with Spina Bifida due to difficulties measuring height and body composition. Instead, show the child and parents the trajectory of the child's weight and height (or other measures of growth and adiposity) on a growth chart as a visual aid. Do not refer to growth cut-offs developed for typically developing children. A steeply increasing trajectory would indicate that overweight or obesity may be a concern and warrant preventative strategies.

- Avoid using scare tactics in older children with Spina Bifida. Instead, discuss the following potential negative consequences of gaining excessive weight, as it relates to their individual circumstances:
 - Moving and transferring may become more difficult, which may also reduce independence and self-care activities.

- Increased pressure on the skin when seated for long periods of time (such as when using a wheelchair) may result in skin breakdown

- Weight gain alongside existing scoliosis or kyphosis may result in additional breathing problems.

- Refer clients to National Center on Health, Physical Activity, and Disability (<http://www.nchpad.org>), which provides advice on nutrition and physical activity for persons with disabilities, including Spina Bifida.

- Consider referral to a "Healthy Lifestyle" program and/or use a mobile application, while recognizing that few such programs are tailored to individuals with disabilities (clinical consensus).

5. Consider the broader literature for all older children, given that there is little evidence that specifically refers teens with Spina Bifida. For instance:

- Understand that eating habits generally worsen as children move into the teen years and become more autonomous.

- Emphasize the positive health benefits of breakfast and eating fruits and vegetables. Skipping breakfast and low fruit and vegetable consumption is common in teens.

- Consider that food insecurity and lower socioeconomic status can be related to poorer diets.

- Emphasize that the family setting remains important for teens. Parental modelling, dietary intake, and encouragement are all associated with fruit and vegetable consumption among teens.

6. Discuss opportunities for the older child to participate in nutrition-related activities, such as:

- Identify the teen's knowledge level about healthy eating habits. (clinical consensus)

- Encourage the family to identify roles that the older child can play as part of daily life, such as in meal planning, shopping, and food preparation.

- Encourage older children to select a new healthy food to try, which can encourage broader food preferences.

- Identify the older child's existing strengths and resources regarding nutrition and how they can be built upon to reach their goals.

7. Screening for diabetes (fasting glucose, HbA1c or oral glucose tolerance test) every two years with a Body Mass Index (BMI) over the ≥85th percentile and have two or more additional risk factors including:

- family history of type 2 diabetes mellitus (T2DM) in a first- or second-degree relative
- high-risk ethnicity
- acanthosis nigricans
- hypertension
- dyslipidemia
- polycystic ovary syndrome (PCOS)

8. Screening for dyslipidemia (fasting lipid profile) is recommended every two years for children with a Body Mass Index (BMI) in the ≥ 85 th percentile or other risk factors for cardiovascular disease (family history of dyslipidemia/early cardiovascular disease and/or morbidity in first- or second-degree relatives, history of diabetes, hypertension, or smoking in childhood).

9. Provide guidance on maintaining good bowel health.

- Explain that increased fiber in the child's diet will add bulk to the stool and make it easier to pass. Sources of fiber include fruit, vegetables, and wholemeal or whole grain bread and cereals.

- Recommend the same guidelines for daily fiber intake that are recommended for all children:³⁴

- 1-3 years: 19g ▪ 4-8 years: 25g
- 9-13 years: female–26g, male–31g
- 14-18 years: female–26g, male–38g

- Recommend that if the child is constipated, parents should increase fiber intake slowly over two to three weeks by adding one new high fiber food every two to three days. Increasing fiber too quickly can make the constipation worse or cause gas, cramping, and diarrhea. (clinical consensus)

- Recommend more fluids, especially water and non-caloric fluids, which will also soften the stool and help with constipation. Follow the following daily maintenance fluid requirements (24-hour period) calculation:

- 100 mL/kg for the first 10 kg body weight
- + 50 mL/kg for the next 10 kg body weight
- + 20 mL for every kilogram of body weight over 20 kg

- Further guidance can be found in the Bowel Function and Care Guidelines.

Sleep Related Breathing Disorders Guidelines

1. Use a standardized sleep questionnaire to query patients at each visit (at least annually) because patients are unlikely to discuss sleep-related symptoms spontaneously with a primary care or specialty provider.

2. Recognize clinical findings that may either contribute to or be the result of sleep disordered breathing: hypertension, obesity, and scoliosis.

3. Improve patients' awareness of SRBD, its presentation and its adverse impact on quality of life.