

# Guidelines for the Care of People with Spina Bifida 6-12 years 11 months

## **Care Coordination Guidelines**

1. It is recommended that the Spina Bifida Care Coordinator provide developmentally-appropriate care education across spectrum of symptoms and conditions related to Spina Bifida to better empower children and their families to manage their own care and be able to recognize complications and emergencies. Identify and/or improve gaps in the family knowledge base specifically related to the school age period (mobility progress, skin inspection, bowel and bladder care, academic/cognitive development, school and social functioning, 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 primary school functioning and update school education and health plans. Encourage participation in age-appropriate activities outside of school with peers, with and without Spina Bifida. Encourage participation in activities such as camps or special family weekends that provide safe places to develop peer relationships with children who may have similar medical challenges. (clinical consensus)  
  
(Appendix: Early Intervention Services, Individualized Educational Plans (IEP) and 504 Plans)
3. It is recommended that the Spina Bifida Care Coordinator coordinate with the family and multidisciplinary Spina Bifida care team to ensure that the child 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 assess family dynamics in coping with the diagnosis and evaluate psychosocial stressors for the family. Assess for depression and anxiety and assist with referrals to mental health and social services when appropriate. (clinical consensus) (Mental Health Guidelines)
5. It is recommended that the Spina Bifida Care Coordinator should work with the child, his or her family and Spina Bifida team members and therapists to start progress on self-management goals and education. Monitor family progress in self-management at regular intervals and clinic visits. Engage the school nurse to help facilitate self-management and independence.<sup>14</sup> Teach self-advocacy and encourage the child to participate as much as possible in his or her own self-management.<sup>15</sup> (Self-Management and Independence Guidelines)
6. When applicable, 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 Spina Bifida Care Coordinator should use two-way communications to identify and address medical concerns and obtain updated records from the primary care provider and/or medical home such as immunizations, growth charts, developmental screenings, and other materials.

7. 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 child with Spina Bifida and monitor family needs and prescriptions for durable medical equipment, supplies, and medications, as needed.

### **Health Promotion and Preventive Health Care Services Guidelines**

1. Monitor that the child is making routine well-child visits to their primary care provider to receive age-appropriate health promotion and preventive services, including age-appropriate screenings for: (clinical consensus)
  - o Hypertension. Since there is no agreement on blood pressure targets for patients with Spina Bifida, it is recommended that baseline blood pressure be monitored to know what is considered hypertensive for a 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, and depression. (Mental Health Guidelines)
- o Motor vehicle and wheelchair safety. (Mobility 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. (Physical Activity Guidelines)
5. Provide information about accessible physical activity and recreational opportunities in the community. (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 specifically-intended to monitor Spina Bifida conditions as well during well-child visits. (Bowel Function and Care Guidelines, Mobility Guidelines, Neurosurgery Guidelines, Orthopedics Guidelines, Skin Care Guidelines, Urology 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. (Sleep Related Breathing Disorders Guidelines)
  - 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 and 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 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 unique neuropsychological issues in transition and self-management. (Neuropsychology Guidelines, Self-Management and Independence Guidelines)
3. Provide updates for families on information regarding long-term financial, insurance, and supportive living planning based on the child's probable trajectory into adult function.
4. Review expectations where individuals with Spina Bifida can access comprehensive care throughout the lifespan including chronic condition management, preventative care, and transition care.
5. Review the clinic's transition policy with patients and families at age 12

### **Family Functioning Guidelines**

1. Provide support and ongoing counseling for parents, the child, and siblings, as needed.

2. Provide anticipatory guidance for parents regarding possible behavioral challenges and autonomy needs in children with Spina Bifida and their siblings. (Mental Health Guidelines, Neuropsychology Guidelines)
3. 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).
4. Assess family dynamics and relationships with school staff. (clinical consensus)
5. Have detailed discussions about appropriate interventions to address academic and social difficulties. Provide parents with current and accurate information about various school settings. For each type of setting, identify potential gaps and determine the impact that such a setting has on family members and the family system. (clinical consensus)
6. Encourage advocacy activities and resources and motivate parents to advocate for themselves and their children with medical, educational, and agency staff. (clinical consensus)
7. Advise parents of their child's 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)
8. Serve as a resource to school systems regarding health issues, individualized educational planning [INSERT LINK TO IEP/504], and socialization. (clinical consensus)
9. Reinforce appropriate family leisure activities. (clinical consensus)
10. Reinforce effective parental discipline, behavioral management, and expectations.
11. Encourage the family to facilitate medical self-management in their children with Spina Bifida, as developmentally appropriate. (Self-Management and Independence Guidelines)
12. Facilitate parents' understanding of the importance of fostering their child's independence and participating in chores and other activities of daily living. (Self-Management and Independence Guidelines)
13. Encourage social activities such as sleepovers, camp overnights, dating, and social and recreational activities outside the home. Encourage development and maintenance of friendships.
14. Emphasize positive attitudes, assertiveness, and self-empowerment of family members. (clinical consensus)
15. Encourage the family to develop strategies that gradually empower their children toward independence such as decision-making and problem-solving. (Self-Management and Independence Guidelines)
16. 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. Encourage participation in community activities for recreation. (Physical Activity Guidelines)
2. Promote the development of friendships by helping families to identify social opportunities (e.g., participation in camps, adaptive sports programs/events, Walk-N-Roll for Spina Bifida, Boy

and Girl Scouts, church youth groups, YMCA activities, and SBA and SBA Chapter social events).

3. Assess the child for depression, anxiety, bullying (including cyber bullying), and social participation. Similarly, identify the child's strengths and build on resources that encourage resilience. Initiate individual and family interventions when appropriate.

4. Encourage activities and hobbies that improve face-to-face social contact. (clinical consensus)

5. Promote transfer of age-appropriate medical responsibility from parent to child in those who have the requisite abilities and cognitive capacity. (Family Functioning Guidelines)

6. Discuss the importance of increasing household responsibilities that are appropriately modified to account for mobility and cognitive limitations. (clinical consensus)

7. Refer children with emotional and/or behavioral difficulties for psychological support and counseling. Identify community resources for social and psychological development (e.g., camps, recreation centers and more).

8. Assess the family's relationship with their child's school and encourage parents to be advocates for their children in the school setting. (clinical consensus) (Family Functioning Guidelines)

9. Promote the child's independence and choice in social activities. Promote self-care so that the child is able to be independent in social settings. (Self-Management and Independence Guidelines)

10. Promote appropriate after-school sports and club activities.

11. Provide additional age-appropriate information/knowledge about Spina Bifida as the child grows. Begin to include child in clinical decision-making. (Neuropsychology Guidelines)

12. Promote and encourage participation in community and SBA and SBA Chapter-related activities. (clinical consensus) (<http://spinabifidaassociation.org/chapters/>)

(<http://spinabifidaassociation.org/national-resource-directory/>)

## **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)

2. Consider strategies to optimize peer relationships. (Mental Health Guidelines)

3. Consider strategies to assess and strengthen family functioning, which can be of critical importance in QOL outcomes in children. (Family Functioning Guidelines)

4. Refer to community resources that enhance protective factors, such as sports, camps, scouts, and other community programs. (Self-Management and Independence Guidelines)

5. Address assessment of executive function. (Neuropsychology Guidelines)

### **Continence**

1. Target strategies to optimize bowel program effectiveness as any bowel incontinence has the greatest negative impact on (Bowel Function and Care Guidelines)

2. Assess both volume and frequency of urinary incontinence, as volume may be more distressing than frequency. (Urology 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 of QOL**

1. Use a systematic approach to evaluating QOL/HRQOL.
2. Consider using both self and parent-report instruments.
3. If feasible, use Spina Bifida and age-specific HRQOLs instruments that measure perception (“concerned about,” “worried about,” “avoid”) and avoids the problem of focusing on function in the physical domain (walking long distances, climbing stairs, jumping) when assessing children with Spina Bifida. Omit any measure that captures the impact in the physical domain. Emotional, social, and school/cognitive domains in most perception-based instruments are useful. (Appendix 1)
4. Consider using a single-item QOL question such as “How would you rate your quality of life?” on a scale of 0-100 with 0=poor and 100=excellent? (Appendix 1) Individual and family factors associated with HRQOL in adolescents and young adults with Spina Bifida should be explored with follow up assessment if needed.

### **Self Management and Independence Guidelines**

1. Provide instruction and support to children and families regarding the knowledge and skills needed to manage Spina Bifida and related independence issues. Teach the child basic self-management skills, including skills to prevent secondary conditions (clean intermittent catheterization, skin care, equipment care, bowel and bladder care, wheel chair maintenance and propulsion) based on individual abilities. Focus on self-efficacy. 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. (Neuropsychology Guidelines)
2. Assist families in learning how to incrementally involve the child in organizing school work and self-management activities and how to begin to transition from parents doing to child doing with parental oversight to eventually child doing without parent oversight.
3. Discuss the need to expand the range of daily life activities and chores as well as strategies to accommodate the child’s learning style and/or mobility.
4. 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. (Mental Health Guidelines, Neuropsychology Guidelines)

7. Assess for potential patient, family, or environmental barriers to developing autonomy and independence, including family stress and conflict, and address in action plan. (Family Functioning Guidelines)
8. Assess bladder and bowel management programs for eventual independent self-management (Bowel Function and Care Guidelines, Urology Guidelines).
9. Consider using an age-and condition-appropriate assessment instrument (Appendix A) especially if the child has executive-functioning impairments.
10. Discuss with parents the need to help their child develop basic money management skills. If the child has an Individualized Educational Plan (IEP), encourage parents and the school to include money management skills in the child's IEP. (clinical consensus)
11. Encourage families to facilitate their child's language performance by creating intellectually- and culturally-enhancing activities in the child's typical environment.
12. Set beginning expectations for independent living. (clinical consensus)
13. Encourage use of technology to enhance self-management. (clinical consensus)

### **Neuropsychology Guidelines**

1. Orient health care professionals that an individual with SBM does not simply have an orthopedic impairment. Explain to them that brain malformations and hydrocephalus (with or without shunting) affect learning, especially in areas that require the construction and integration of information such as self-management skills. Learning is facilitated when it is based on rules that can be verbally mediated and rehearsed, much like a recipe. This is especially important for bladder and bowel interventions for which the child's participation at an early age facilitates independence and social adjustment and adherence to dietary regimens. Abstract concepts and global guidelines about self-care are ineffective for skill acquisition. It is essential to create routines, so that practice and repetition of self-management tasks can become rote activities. Coach clinical teams to carefully formulate clinical instructions to be verbally mediated and to emphasize rule-based learning with repetition and rehearsal. (clinical consensus) (Health Promotion and Preventive Services Guidelines, Nutrition and Obesity Guidelines)
2. Orient educators and school-based professionals that an individual with SBM does not simply have an orthopedic impairment and that brain malformations and hydrocephalus (with or without shunting) affect learning, especially in areas that require the construction and integration of information such as language, reading comprehension, and math problem-solving. Psycho-educational assessments can track global intellectual and academic progression, but rarely assess the development of essential skills in attention, executive functioning, coordinated upper limb, and memory domains, as well as adaptive skill acquisition. Children with SBM benefit from a full neuropsychological assessment, when available. (clinical consensus)
3. Monitor school age children carefully for the onset of academic, attention, and behavioral difficulties. These problems tend to be identified later in school, partly because of the early development of word recognition, rote numerical skills, and vocabulary skills (usually in children who are not from socially- and economically- disadvantaged settings) that mask the presence of difficulties with math and reading comprehension. (clinical consensus)
4. Carefully monitor children for the onset of attention problems, as they are often interpreted as motivational or behavioral issues and are often manifested as lack of focus, slow cognitive

tempo, failure to initiate, and infrequently with hyperactivity or impulsivity.<sup>23</sup> Attention problems are correlated with the Chiari malformation, tectal beaking, and hypogenesis of the corpus callosum

5. Follow American Academy of Pediatrics guidelines when evaluating for ADHD. One-third of individuals meet criteria for ADHD, predominantly Inattentive Type on parent rating scales. Interventions for attention problems that involve medications may be tried, but clinical experience suggests that lower doses are effective and that many children with SBM do not respond robustly to stimulants, most likely because the underlying attention problem emerges from posterior components of the attention network and not from the frontal-striatal networks (as in developmental ADHD). (clinical consensus)

6. Monitor children for the development of language and reading problems. The severity of hydrocephalus and corpus callosum malformations affects the child's ability to integrate information and to construct meaning from language. Over 25% of children with SBM have significant language and reading comprehension problems, which tend to be present both for listening and reading comprehension. Because of these common academic difficulties in children with SBM, formal assessment should include text-level reading comprehension and not just word reading accuracy and fluency.

7. Monitor children for the development of math problems. Over 50% of children with SBM develop math difficulties. Assessment of mathematics should include assessment of complex calculation skills and, in the later grade school years, math word problems.

8. Implement interventions like those used with children with learning disabilities when a child has a problem with reading or math, as these are often effective. For example, although problems with word reading and phonological awareness are rare in children with SBM, treatment programs like those used with children with dyslexia have been shown to be effective. Another example is the successful use of math problem-solving interventions designed for children with math disabilities. Take advantage of children's strengths in rule-based learning by providing explicit, well-structured instruction.

9. Use assistive devices as early as possible when developing writing programs. Keyboarding is a viable alternative to handwriting, although some practice with paper and pencil skills is useful through most of elementary school. Keyboarding must be taught and rehearsed if it is to be useful. Accommodations for writing difficulties are critical components of the educational plan. (clinical consensus)

10. When available, consider full neuropsychological evaluations that include the assessment of early literacy and numeracy skills. Neuropsychological assessments provide a more comprehensive understanding of strength and weakness, as well as significant discrepancies that may not be captured by psycho-educational testing that is provided by school districts. (clinical consensus)

## **Neurosurgery Guidelines**

**Patient/Family** 1. Continue to encourage the family to observe the child for clinical signs of shunt failure, brainstem dysfunction, TSC and syringomyelia. (clinical consensus)

2. Foster and develop working relationship with the team of Spina Bifida providers. (clinical consensus)

3. Motivate the family to establish working relationships with their child's educational system including teachers and other educational professionals. (clinical consensus)
4. Urge the family to collaborate with the clinic coordinator and/or social worker to optimize resources in the setting of potential neurocognitive dysfunction, and to identify and relay neurocognitive changes to the medical team. (clinical consensus) (Neuropsychiatry Guidelines)

#### **Providers/Neurosurgeons/Spina Bifida Clinic**

1. Follow children ages 6-12 years 11 months at 12-month intervals in the Spina Bifida clinic. (clinical consensus)
2. Review the signs of acute shunt failure (headache, neck pain, vomiting, and lethargy/sleepiness), and chronic shunt failure (recurring low grade headache and neck pain; loss of developmental milestones; cognitive, behavioral, or neurological decline; and orthopedic or urological regression) with the family. Follow the child clinically to observe for these signs.
3. Teach or review with the family and urge them to observe for the signs of TSC (back pain, declining lower extremity sensorimotor function, bladder or bowel control decline and progressive orthopedic deformities and/or scoliosis). Follow the child clinically to observe for these signs.
4. Teach or review with the family and urge them to observe for signs of syringomyelia (neck or back pain and sensorimotor changes in arms and hands). Follow clinically to observe for these signs. (clinical consensus)
5. Review the signs of brain stem dysfunction that might occur in this age range (poor control of secretions, swallowing dysfunction, stridor, and declining language function) with the family. Follow clinically to observe for these signs. (clinical consensus)
6. To augment clinical decision-making, use adjunctive studies during routine visits with the well child (for example, imaging such as MRI/CT and urodynamic and sleep and swallow studies), doing so judiciously and according to experience, preference, and best clinical judgment. (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. Discuss with families the benefits of the different types of mobility devices including ambulation aides and wheelchairs based on predicted mobility potential.
3. Monitor walking or wheeling ability with standardized outcome measures. Consider gait studies if ambulation is changing or information is needed on optimizing bracing.
4. Continue flexibility, range of motion (ROM) and strengthening exercises to maintain mobility goals, whether using ambulation devices or a wheelchair.
5. Teach independence in putting on and taking off orthoses. (clinical consensus)
6. Educate child about importance of physical activity to maintain flexibility, strength and health, especially during growth years and explore adapted physical education opportunities or recreational sports options with the family. (Physical Activity Guidelines)

7. Start teaching children to be involved in their own care by educating them to watch for signs and symptoms of pressure injuries, fracture, and neurologic changes. (clinical consensus) (Self-Management and Independence Guidelines)
8. Ensure proper wheelchair fit, posture, and technique in children who use wheelchairs, in order to reduce energy expenditure and promote long-term function.
9. Encourage weight-bearing activities every day to promote bone health. (clinical consensus)
10. Collaborate with orthopedic specialists to monitor for age-specific musculoskeletal problems. (Orthopedic Guidelines)

### **Orthopedics Guidelines**

1. Monitor gait, rotational deformities and foot position. (clinical consensus)
2. Consider correction of foot deformities to facilitate orthotic management with soft tissue release, tendon transfer and osteotomy, if necessary. It is recommended that fusion be avoided if possible. (clinical consensus)
3. Consider correction of tibial and femoral rotational deformities when they are interfering with gait and precluding orthotic management.
4. Consider conducting computerized gait analysis, when available, in children with low lumbar or sacral level lesions who have atypical gait abnormalities. This information will be helpful when making decisions regarding surgery or bracing.
5. Monitor for the development of scoliosis/kyphosis. (clinical consensus)
6. Obtain anteroposterior and lateral scoliosis radiographs every one to two years if deformity is suspected clinically. Do so more frequently in patients with progressive spinal deformity. Perform radiographs in a sitting position in those who can sit but not stand and in a standing position in patients who can stand. (clinical consensus)
7. It is recommended that surgical treatment of scoliosis be reserved for a progressive deformity that is unresponsive to non-operative management. An example is when the scoliosis has progressed in spite of bracing and after a neurosurgical cause, such as a tethered cord, has been ruled out. It is also recommended that management with growing rod surgery and fusionless technique should include spinal cord monitoring in children with distal neurologic function. Growing rod surgery with sacral-pelvic fixation is effective in correcting deformity and achieving growth.
8. Consider surgical treatment of gibbus deformity for intractable skin breakdown or to free up the upper limbs for independent sitting. The current literature describes multiple techniques.
9. Teach children and families about fractures and related precautions. (clinical consensus)

### **Physical Activity Guidelines**

1. Discuss the benefits of participating in physical activity, recreation, and sports with children with Spina Bifida.<sup>24,29</sup> Discuss with parents/caregivers the importance of limiting sedentary behaviors.<sup>20</sup> Encourage parents to give their child 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 parents/caregivers follow the National Physical Activity Guidelines for their child with Spina Bifida 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.
  - 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 the child and parents/caregivers to talk about physical activity goals and work through barriers. Support parents/caregivers to develop an action plan with strategies to support their child's participation in physical activity in their community and school. Prescribe, using a prescription pad, physical activity based on goals identified by the child. (clinical consensus)
4. Perform pre-participation evaluations for children with Spina Bifida in collaboration with the child and family, pediatric specialists, therapists, coaches, and others to identify medical risks and modifications that can be made to ensure participation.
5. Identify strategies to minimize risk of illness and injury related to participation through activity adaptations and safety precautions. Identify and provide additional support and information for 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.
6. 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)
7. 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.
8. Advocate for and address barriers to participation of children with Spina Bifida in physical activity, recreation, and sports.
9. Inform parents/caregivers of their child's right to adapted physical education/activity and encourage parents/caregivers to advocate for physical activity goals to be added to their child's IEP or Section 504 plan (if eligible for IEP or Section 504 plan).
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.

### **Men's Health Guidelines**

1. Provide anticipatory guidance regarding sexual function and its potential challenges. (clinical consensus)

2. Conduct an annual scrotal exam that documents testicular position, size, consistency, symmetry, and presence or absence of masses. (clinical consensus)
3. Access and document genital sensation (penile, scrotal) and Tanner staging annually.
4. Offer human papillomavirus (HPV) vaccination per Centers for Disease Control and Prevention and American Academy of Pediatrics guidelines, if appropriate.

### **Sexual Health and Education Guidelines**

1. Provide factual information to parents and caregivers and encourage them to provide developmentally-appropriate sexual education to their children.
2. Review relevant literature that addresses sexual health and education, such as “Bright Futures” and other reports prepared by the American Academy of Pediatrics.
3. Allow the child to ask questions about sexual development and sexuality.
4. Serve as a resource to schools to ensure that children with Spina Bifida participate in sexual education.
5. Encourage parents to discuss information that their children are receiving about healthy relationships from school, their peers, the media, and social media.
6. Promote skill-building to identify dangerous situations, refuse or break off a sexual attack, and summon help.
7. Promote socially-appropriate behaviors and social skills.
8. Underscore the goal of continence (Bowel Function and Care Guidelines, Urology Guidelines) for optimal sexual relationships in the future (clinical consensus)
9. Provide education about sexuality, pubertal development, evaluate concerns or abnormal physical findings, and explain the risks of precocious puberty (Endocrine: Puberty and Precocious Puberty 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 recurrent symptomatic UTIs or if urodynamic testing identifies bladder hostility. (clinical consensus)
3. Obtain urodynamic testing when initiating a urinary continence program, if the following are present: (clinical consensus)
  - upper urinary tract changes such as hydronephrosis or renal scarring
  - recurring symptomatic UTIs
  - changes in urinary continence status
4. Obtain a serum creatinine test yearly. If the child has low muscle mass, consider an alternative measure of renal function. (clinical consensus)
5. Obtain serum chemistries yearly on any child who has had urinary reconstruction.
6. Obtain a serum B12 level test every year beginning two years after urinary reconstruction. (clinical consensus)
7. Discuss a urinary continence program and interest in beginning the program and options at each visit. (clinical consensus) (Self-Management and Independence Guidelines)
8. Discuss a bowel management program and the interest and options at each visit. (clinical consensus) (Bowel Function and Care Guidelines)

## **Women's Health Guidelines**

1. Puberty occurs earlier in girls with Spina Bifida than in the general population. It is recommended that, along with Tanner staging, care providers discuss the possibility of early puberty with girls and their families and create an atmosphere of open communication. (Endocrine: Puberty and Precocious Puberty Guidelines, Sexual Health and Education Guidelines)
2. Offer human papillomavirus (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 the goal of bowel continence and institute 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 twofold attack of oral and rectal interventions to meet the goal of bowel continence without constipation or fecal incontinence.
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 MACE.
10. Refer to a Spina Bifida clinic or specialist with expertise in bowel management in Spina Bifida. (clinical consensus)

## **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 same 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 abnormal signs of puberty are observed. Abnormal signs could include progressive breast development over a 4- to 6-month period of observation or progressive penis and testicular enlargement, especially if accompanied by rapid linear growth. Children exhibiting these true indicators of early puberty need prompt evaluation by an appropriate pediatric endocrinologist.
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)

## **Endocrine: Short Stature and the Effects of the Human Growth Hormone**

### **Guidelines**

1. Assess weight, height at each health supervision visit. If height is not able to be measured using a stadiometer, it is recommended that a consistent parameter (such as arm span) should be measured and recorded. (clinical consensus)
2. Have a discussion with the family about the expected height of the child, based on the limitations due to myelomeningocele and the parents' height. (clinical consensus)
3. Discuss the risks and benefits of hGH therapy with the parents. (clinical consensus)
4. If concerns about growth arise, a referral to a pediatric endocrinologist is recommended for growth assessment, IGF-1, IGF Binding Protein-3, and GH stimulation tests. (clinical consensus)
5. If hGH treatment is initiated, monitor pituitary function, scoliosis, tethering of spinal cord, growth velocity, and pubertal development. This may be done in collaboration with a pediatric endocrinologist. (clinical consensus)

### **Integument ( Skin) Guidelines**

1. Teach parents and caregivers to inspect the skin daily (especially weight bearing or insensate areas) for changes in color, texture, and temperature.
2. Encourage the child's involvement in skin inspection.
3. Teach child to develop awareness of insensate areas.
4. Review with parents and caregivers the consequences of heat, moisture, or pressure related to insensate areas.
5. Teach parents/caregivers how to inspect for well-fitting orthoses and other equipment that may cause injury to skin.
6. Teach parents and caregivers that the child should wear protective clothing and footwear over insensate areas.
7. Discuss the need to check water temperature and encourage the use of a bath water thermometer.
8. Tell parents and caregivers to check for hot surfaces that have been exposed to the sun such as car seats and playground equipment.
9. Promote adequate hydration and proper nutrition for healthy skin. (Nutrition, Metabolic Syndrome, and Obesity Guidelines)
10. Encourage parents, caregivers, and the child to keep skin clean and dry.
11. Suggest wearing seamless socks that are clean and dry.

12. Suggest the use of antiperspirant on areas with perspiration, such as the feet and intertriginous areas.
13. Tell parents and caregivers to seek treatment if the child's skin is compromised.
14. Advise parents and caregivers to engage non-ambulatory children in pressure-relieving activities every 15 minutes.

### **Latex and Latex Allergy Guidelines**

1. Educate school-age children about their avoidance of latex products such as latex-containing urinary catheters and inform them 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. Tell parents and caregivers of children identified as having a latex allergy, and the children themselves, to have diphenhydramine and self-administered epinephrine available at all times. (clinical consensus)
4. Instruct families to check that food made in public venues has been prepared with latex-free gloves. (clinical consensus)
5. Urge children to continue following latex precautions because risk-taking during the teen years is common. (clinical consensus)
6. Review the principles of latex precaution with the child during a clinic visit and answer any questions. (clinical consensus)
7. Refer to an allergist when the child is allergic to latex but does not know if he or she is allergic to cross-reacting foods; this is particularly crucial in those who have had a systemic or anaphylactic episode. (Appendix 2) 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. 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 assessment 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 and Body Composition Measurements)
3. Conduct annual assessment of blood pressure/percentiles to monitor for pre-hypertension and hypertension. (clinical consensus)
4. Highlight dietary needs specific to living with Spina Bifida.
  - Discuss the importance of consuming fiber and water to manage bowel and bladder health. Sources of fiber include fruit, vegetables, whole wheat or whole grain bread and cereals. A mix of each along with regular fluids will help avoid constipation.
  - Advise limiting sugary drinks such as juice, chocolate milk, and sports beverages.
  - Discuss that children with Chiari malformation may have a sensitivity to different food textures.

Recommend that the child have access to food purchasing and preparation spaces.  
(clinical consensus)

- Recommend a diet with adequate calcium and vitamin D for children with Spina Bifida, in order to avoid fractures due to osteoporosis.

- Consider that adults and families with lower incomes may experience food insecurity.

- 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.

5. Provide families with nutritional information tailored to their circumstances.

- Take into account a family's geographical location, ethnicity, access to food, and other related factors when providing dietary education.

- Consider that adults and families with lower incomes may experience food insecurity.

- Encourage parents to include their children from an early age to participate in grocery shopping and food preparation, as appropriate to their age and ability.

- Suggest parents to let their children choose a new healthy food to try. Involving children in choices in food selection can lead them to increased independence and interest in their foods and to learn about making healthy choices. Repeating their exposure to healthy food options can increase children's acceptance and enjoyment of these foods.

- Involve children in discussions about healthy lifestyles in order to explore their understanding, perceptions, and priorities regarding nutrition.<sup>28</sup> If appropriate, ask parents to identify one or two small, healthy nutrition changes that they feel they could integrate into their daily life.

- Consider making a referral to a healthy lifestyle program and/or a smartphone application, while recognizing that few such programs are tailored to individuals with disabilities.  
(clinical consensus)

- Celebrate any successes, such as drinking more water, introducing a new fruit or vegetable, cutting back on sugary drinks, and having regular meal times. Focus upon the strengths of the family.

- Highlight the importance of parents modeling healthy behaviors themselves to their children from an early age. Encourage the whole family to get involved in healthy living activities not just the child with Spina Bifida

- Understand that experiencing food insecurity may lead to a poor-quality diet, and have developmental consequences on the child.

- Highlight that children with Spina Bifida, especially those who are non-ambulatory, undertake low levels of physical activity, and have higher body fat levels or contractures, are at increased risk for bone fractures. Recommend a diet with adequate calcium and vitamin D.

6. Screening for diabetes (fasting glucose, HbA1c or oral glucose tolerance test) is recommended every two years in children 10 years of age or older (or at the onset of puberty if it occurs at a younger age), and for all children with a Body Mass Index (BMI) over the ≥85th percentile and who 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)

7. Screening for dyslipidemia (fasting lipid profile) is recommended every two years for children up to 8 years of age with a Body Mass Index (BMI) over the 95th percentile or other risk factors for cardiovascular disease, such as:

- family history of dyslipidemia/early cardiovascular disease and/or morbidity in first- or second-degree relatives
- history of diabetes, hypertension, or smoking in childhood

8. Screening for dyslipidemia (fasting lipid profile) is recommended once for all children ages 9-11 years.

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 24-hour period daily maintenance fluid requirements 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. Recognize that the symptoms of SRBD in children (mouth breathing, a history of delayed growth, features of inattention and hyperactivity) are different compared to adults (snoring and excessive daytime sleepiness are less frequent).

2. Ask questions related to sleep quality, quantity and other possible symptoms at every visit (at least annually). Standardized screening questionnaires for SRBD in children are useful in clinical settings.

3. Further evaluate changes in respiratory status/function.

4. Discuss sleep disordered breathing with parents and care providers so they can better observe for early symptoms or changes.