

Guidelines for the Care of People with Spina Bifida

18+ years

Care Coordination Guidelines

1. It is recommended that the Spina Bifida Care Coordinator be knowledgeable about the resources for adults with Spina Bifida in their geographic area and coordinate the successful transition from pediatric to adult providers for adults with Spina Bifida. Where appropriate, educate adults with Spina Bifida about the key differences between adult and pediatric providers, including the possibility that there may not be an adult multidisciplinary Spina Bifida team nearby. Inform adults with Spina Bifida on the importance of having a primary care provider. (Transition Guidelines)
2. It is recommended that the Spina Bifida Care Coordinator conduct an inventory of the adult's ability to provide self-management, complete activities of daily living, and manage mobility equipment and transportation needs. For those who have a significant intellectual disability and may be unable to live independently, assist the family with the conservatorship process and with maintaining Supplemental Security Insurance (SSI) and other types of insurance coverage. (clinical consensus) (Self-Management and Independence Guidelines)
3. It is recommended that the Spina Bifida Care Coordinator assist with referrals to vocational and employment training opportunities, as needed. (clinical consensus)
4. It is recommended that the Spina Bifida Care Coordinator assist with referrals to driver education training opportunities, as needed and/or referral for appropriate pre-driver training evaluations including vision, ability to use lower extremities vs. hand controls and other driving requirements. (clinical consensus)
5. It is recommended that the Spina Bifida Care Coordinator coordinate with Spina Bifida care providers to determine if the person is up-to-date on all sub-specialty care visits, imaging and monitoring and equipment needs, where appropriate. This may include assistance with insurance authorization. (Mobility Guidelines, Neurosurgery Guidelines, Orthopedics Guidelines, and Urology Guidelines)
6. It is recommended that the Spina Bifida Care Coordinator assess individual dynamics in coping with living with Spina Bifida and evaluate psychosocial stressors for the individual. 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)
7. It is recommended that the Spina Bifida Care Coordinator should serve as the lead contact person and information provider for the Spina Bifida clinic and monitor individual needs and prescriptions for durable medical equipment, supplies, and medications as needed. Special considerations may be needed to apply care coordination principles to assist adults who see multiple providers independently.
8. It is recommended that the Spina Bifida Care Coordinator assess and monitor for clinical deterioration, loss of mobility, chronic pain, obesity, and use two-way communication between the Spina Bifida Care Coordinator and the primary care provider and/or medical home to assess

and address concerns and assist with medical referrals, as appropriate.17,20 (Health Promotion and Preventive Medicine Guidelines, Mobility Guidelines, Neurosurgery Guidelines, Nutrition, Metabolic Syndrome, and Obesity Guidelines, Orthopedics Guidelines, Urology Guidelines)

Health Promotion and Preventive Health Care Services Guidelines

1. Monitor that the adult is receiving typical and age-related health promotion and preventive services, including screening or counseling about:

- 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 the particular child.

- o Lipid disorders. Treat as needed.

- o Overweight/obesity. Counsel as to healthy diet and exercise habits. (Nutrition, Metabolic Syndrome, and Obesity Guidelines)
- o Cancer, including skin cancer. Promote age-appropriate screening.

- o Diabetes and metabolic syndrome. Screening and treatment as needed.

- o Fall prevention.

- o Adaptive physical activity. (Physical Activity Guidelines)

- o Depression and anxiety. (Mental Health Guidelines)

- o Smoking and illicit drug use.

- o Hearing and vision.

2. Monitor for comorbid conditions that are specific to adults with Spina Bifida, both during visits that are specifically intended to monitor Spina Bifida conditions as well as routine visits to their PCP. (Bowel Function and Care Guidelines, Mobility Guidelines, Neurosurgery Guidelines, Orthopedics Guidelines, Skin Care Guidelines, Urology Guidelines) Check for:

- o Shunt concerns. Monitor for neurologic changes.

- o Sleep apnea. May need a referral for pulmonary evaluation and sleep study.

- o Skeletal and limb deformity. Check for new problems with positioning or brace use and new pain.

- o Osteoporosis. Counsel about the need for weight-bearing activities.

- o Pain. Use age- and cognition-appropriate pain scale to assess.

- 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 for changes in bowel/bladder function³ (Bowel Function and Care Guidelines, Urology Guidelines)

- o Skin breakdown and pressure injury. Encourage adults to conduct frequent skin checks and to shift their weight at least every two hours.

- o Lymphedema.

- o Adaptive equipment needs such as for orthoses, crutches, walkers, and wheelchairs. Make referrals to necessary subspecialists.
- 3. Promote care coordination between Spina Bifida-specific subspecialists and primary care providers. (Care Coordination Guidelines)
- 4. Educate adults about early signs of chronic conditions related to Spina Bifida.
- 5. Counsel about and monitor for sexually transmitted infections, use of different types of contraceptives, and violence among intimate partners. (clinical consensus) (Men's Health Guidelines, Sexual Health and Education Guidelines, Women's Health Guidelines)
- 6. Provide counseling about family planning and possible fertility and genetic counseling to individuals interested in pregnancy. Recommend counseling about prenatal vitamins and folic acid. (clinical consensus) (Men's Health Guidelines, Women's Health Guidelines)
- 7. Promote self-management for health and health care services. Assess the adult's ability to perform routine care needs such as bowel, bladder, and skin-check regimens, their ability to detect changes in their health status, and their awareness of their need for provider services to maximize their independence. (Self-Management and Independence Guidelines)

Transition Guidelines

1. Provide updates for adults and families regarding a probable trajectory for adult function and expectation 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. Continue to assist with transition coordination as applicable including:
 - a. Assistance in identifying adult providers who accept the patient's insurance and can assume his/her care.
 - b. Counseling regarding long-term financial, insurance, and supportive living plans (housing, transportation, etc.) based on the individual's current needs and probable trajectory of adult function.
 - c. Information for education/employment transition support as applicable to the individual's need such as vocational rehabilitation services, school transition planning, as part of the Individualized Educational Plan [INSERT LINK], and adaptive vocational needs.
 - d. Decision-making supports and modalities that maximize the individual's ability to participate in decisions for themselves, such as a medical power of attorney, supportive decision-making, or guardianship. Referral to medical legal partnerships may be needed.
 - e. Adult disability determination information, if applicable.
 - f. Creation of a medical summary including past medical and surgical history, current care plans, medications, allergies, vaccines, and current providers.

g. Self-management support. (Self-Management and Independence Guidelines)

Consider using transition and self-management assessment tools to direct goals and interventions.

4. Ensure that patient-centered and developmentally appropriate preventive and chronic condition management services are provided throughout transition. 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, young adults, and siblings, as well as older adults with Spina Bifida, as needed.

2. Work with families to support the development of maximal vocational and social independence.

3. Continue to work with the family to support medical self-management in their young adult.

4. Continue working with the family to ensure a successful transition to adult health care. (Transition Guidelines)

5. Work with the young and older adults to navigate sexual expression in a safe and mature fashion. (Sexual Health and Education Guidelines)

6. Assess the family context for helping the young adult 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. Screen for depression or anxiety and initiate interventions when appropriate.

2. Continue the transfer of medical responsibilities in young adults with Spina Bifida who have the requisite abilities and cognitive capacity.

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

4. Encourage ongoing efforts to promote friendship and social intimacy.

5. Encourage and promote vocational or occupational goals and pursuits. (clinical consensus) (Transition Guidelines)

6. Maintain efforts for good general health promotion and exercise, as well as specialized Spina Bifida care. Optimize health to reduce the risk of obesity and maximize social opportunities and mental health. (Physical Activity Guidelines)

7. Recommend SBA resources (<http://spinabifidaassociation.org/learn-about-sb/adults/>). (clinical consensus)

8. Continue to refine the plan to ease transition from pediatric to adult health care. (Transition Guidelines)

Quality of Life Guidelines

Psychosocial well-being

1. Identify strategies or resources to facilitate the development of protective beliefs (e.g. hope, optimism, attitudes, future expectations, active coping strategies) and behaviors such as showing affection, bouncing back when things don't go their way, showing interest in learning

new things, handling negative situations, and establishing and maintaining friendships. (clinical consensus) (Mental Health Guidelines, especially the section on peer relationships)

2. Explore satisfaction with relationships and their sexuality. (clinical consensus) (Sexual Health and Education Guidelines)

3. Consider strategies to optimize peer relationships. (clinical consensus) (Mental Health Guidelines)

4. Consider the importance of each individual's QOL unique priorities. (clinical consensus)

5. Refer to community resources such as sports, camps, community advocacy groups, universities with strong programs to support students with disabilities, and other community programs that enhance protective factors. (clinical consensus) (Self-Management and Independence Guidelines)

6. Address strategies to compensate for executive functioning challenges. (clinical consensus) (Neuropsychology Guidelines)

7. Consider strategies to enhance self-management behaviors. (Self-Management and Independence Guidelines)

Continence/mobility

1. Target strategies to optimize bowel program effectiveness as any bowel incontinence has the greatest negative impact on QOL in adults, especially in social domains.

2. Investigate the adult's satisfaction with her/his bowel program. Address concerns to optimize program.

3. Assess both volume and frequency of urinary incontinence in adults, as volume may be more distressing than frequency.

4. Consider functional mobility options that optimize societal participation. (Mobility Guidelines)

Pain

1. Evaluate the 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 an age-and condition-specific instrument to assess HRQOL. Instruments that measures perception ("concerned about," "worried about," "avoid") and avoid the problem of focusing on function in the physical domain (walking long distances, climbing stairs, jumping) are preferred. Omit any measure that captures the impact in the physical domain. Emotional, social, and school/cognitive domains in most perception-based instruments are useful. (Appendix 1). Instruments like the WHOQOL-BREF (Appendix 1) avoid this issue using questions such as "Do you have enough energy for everyday activities?" or "To what extent do you feel that physical pain prevents you from doing what you need to do?" Spina Bifida-and-adult-specific measures also assess perception and avoid this issue.

4. Evaluate both the adult's self-report and the parent report of QOL/HRQOL. If assessment time is limited choose self-report of QOL/HRQOL.. 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 full responsibility for implementing condition-specific self-management behaviors in appropriate areas, as needed (e.g. managing medications, preventing complications, monitoring skin care, maintaining equipment, bowel and bladder care, and ability to make health care appointments).
2. Reinforce the need for daily skin assessment, given the high incidence of skin breakdown on lower extremities (e.g. due to poor fitting leg braces) and risk for wound-related hospitalization. (Integument (Skin) Guidelines)
3. Evaluate if the adult has expanded self-management to encompass everyday living activities such as laundry, meal preparation, managing finances, making health care appointment and ordering supplies.
4. Initiate a discussion and develop an action plans to address deficits in self-management skills, abilities and behaviors, as needed. (clinical consensus)
 - Use a valid and reliable instrument to assess self-management skills, abilities and performance of self-management or independence behaviors over time in adults.
 - Support development of knowledge and skills necessary for self-management (e.g., self-efficacy, decision-making, goal setting, self-regulation, and communication).
 - Evaluate and monitor cognitive functions, as they underpin decision-making and self-management. (Neuropsychology Guidelines)
 - Assess the adult’s ability to use transportation; encourage enrollment in driver’s education (adaptive, if needed) if the adult possesses the necessary cognitive and motor abilities and has not done so already. If driving is not realistic, teach (or encourage the family to teach) the adult how to use transportation (e.g., public transportation, van services for individuals with disabilities, or other transportation options). (clinical consensus)
 - Evaluate the young adult’s ability to live independently and connect with him or her with housing resources, such as Centers for Independent Living. (clinical consensus)
5. Encourage the use of technology in developing basic self-management skills. For instance, using email or a personal online health record, or patient portal to contact the clinic coordinator and physician with questions. Offer alternatives if this form of access is not available or appropriate.
6. Encourage the use of technology programs to enhance self-management outcomes (e.g. using mobile health (mHealth) or telehealth tools to monitor skin breakdown or report response to medication for UTI).
7. Expand the discussion of sexuality, contraception (including latex allergy precautions), marriage, childbearing issues, genetic counseling, and folic acid supplementation. (Sexual Health and Education Guidelines)
8. Expand the discussion on child rearing and parenting issues and resources as appropriate. (clinical consensus)
9. Discuss strategies for safe infant handling (e.g., holding an infant if you use a wheelchair or accessing a crib or car seat) with parents or expectant parents with mobility limitations. (clinical consensus)

10. Encourage involvement in empowerment activities and organizations (e.g., sports, mentoring, camps, and local, national and international Spina Bifida, and other disability organizations).
11. Support family functioning strengths related to self-management including family satisfaction and family resources. (Family Functioning Guidelines)
12. Assess individual and system barriers to self-management (e.g., difficulties with self-advocacy, assertiveness, insufficient adult services).
13. Refer to vocational rehabilitation, independent living centers, or other community agencies as appropriate. (clinical consensus)
14. Provide information about accessible housing, financing, and appropriate outside agencies. (clinical consensus)
15. Encourage planning and use of support services (e.g., in a college setting, services for students with disabilities) for self-management and independence in new environments. (Transition Guidelines)
16. Encourage the use of wellness programs.
17. Evaluate and support patients as their parents and caregivers age and assist individuals with Spina Bifida plan for changes in self-management and independence when their parents and caregivers will not be available. (clinical consensus)

Neuropsychology Guidelines

1. Many patients with intellectual disabilities or significant learning challenges will remain eligible for services through their local school districts until 21 or 22 years of age. When young adults are eligible, these services provide access to both vocational and life skills training that are essential to support the development of stronger functional independence skills. (clinical consensus)
2. Encourage that vocational services addressing job skills, additional education, and related activities be provided to appropriate individuals in a timely manner. Referrals to state-based agencies are commonly included in transition programs, and found in special education documentation/IEPs. (clinical consensus)
3. For students who received special education (IEP) or 504 Plan accommodations in high school, ongoing supports under the Americans with Disabilities Act (ADA)/Section 504 of the Rehabilitation Act are necessary. For those attending college, refer them to their college's office of disability services for ongoing educational supports. Many students will also require an updated neuropsychological assessment to support eligibility. For those in workplace environments, refer to the state-based rehabilitation/vocational commission for additional support. (clinical consensus)
4. In preparation for the transition to adult care models, where often times less coordination of medical care is provided, medical team members must take an active teaching and training role to build the necessary skills to support transition. Teach the person with Spina Bifida the skills necessary to effectively communicate with staff, recognizing that they may prefer a different method than their parents (e.g., phone calls vs. internet portal). Test patients on important aspects of their medical conditions, regimens, and allergies. Rehearse triaging medical symptomology, with clear guidelines on when to seek medical care, to mastery (e.g., not when

they first get it right, but when they always get it right). (clinical consensus) (Self-Management and Independence Guidelines, Transition Guidelines)

5. Continuously monitor cognitive skills, especially math, memory, and attention, to ensure the maintenance of learning skills essential for work and independence. Changes in these areas may be a sign of unidentified shunt failure or shunt dependency, or other significant medical problem requiring intervention. (clinical consensus). Full neuropsychological assessment is recommended for adults with SBM who experience cognitive decline and suspected shunt failure. (clinical consensus)

6. Monitor for mental health concerns and potential cognitive decline with aging. (Mental Health Guidelines)

Neurosurgery Guidelines

Patient/Family

1. Observe the adult for clinical signs of shunt failure, brainstem dysfunction, TSC and syringomyelia. (clinical consensus)

2. Continue fostering a working relationship with the team of Spina Bifida providers. (clinical consensus)

3. Adult and family should be encouraged to review information about transitioning to adult care, including: (Self-Management and Independence Guidelines, Transition Guidelines,)

- Knowledge and autonomy for personal health decisions.
- Awareness of own body symptoms/signs.
- Knowledge about predictors of good quality of life in adulthood.

Providers/Neurosurgeons/Spina Bifida Clinic

1. Follow adults of 18+ years at 12-month intervals in an adult Spina Bifida clinic setting. (clinical consensus)

2. Neurosurgery should assist the patient and family in identifying an adult neurosurgery provider and facilitate and support completion of transitional care. (clinical consensus) (Transition Guidelines)

3. Review with the adult and family the signs of acute shunt failure (headache, neck pain, vomiting, lethargy/sleepiness), and chronic shunt failure (recurring low grade headache/neck pain and changes in behavioral or cognitive function). Follow clinically to observe for these signs. (clinical consensus)

4. Review with the adult and family the signs of brain stem dysfunction in adults (poor control of secretions, swallowing dysfunction, stridor, and declining language function). Follow the adult clinically to observe for these signs. (clinical consensus)

5. Teach or review with the adult and family and urge them to observe for signs of TSC (back pain, declining sensorimotor function, and urologic dysfunction). Follow the adult clinically to observe for these signs

6. Teach or review with the adult and family and urge them to observe for signs of syringomyelia (back pain and sensorimotor changes in arms and hands). Follow the adult clinically to observe for these signs. (clinical consensus)

7. Use adjunctive studies judiciously to augment clinical decision-making (imaging such as MRI/CT, urodynamics, and sleep and swallow studies) during routine visits with the well adult, according to experience, preference, and best clinical judgment. (clinical consensus)
8. Encourage pediatric neurosurgeons to be available for education and teaching opportunities from the adult Spina Bifida team in order to learn how to provide care to adults with Spina Bifida.

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 walking or wheeling ability and check for factors that may negatively impact mobility.
3. Continue to discuss the benefits of being involved in physical activities. (clinical consensus)
4. Continue with home programs to maintain flexibility, range of motion, and strengthening as this will impact mobility. (clinical consensus)
5. Optimize gait with supportive orthoses or devices for balance, monitor for torque forces at the knee or excessive forces in the upper body. (clinical consensus)
6. Teach adults with Spina Bifida about the systems of care related to mobility equipment and orthoses. Adults need to know how to identify who to call when they experience problems with their mobility devices, and the extent of their health insurance coverage and benefits. (clinical consensus)
7. Educate adults on the importance of preventing loss of mobility (both ambulation and wheelchair) through the use of appropriate technique and maintaining a healthy weight and level of strength. (clinical consensus)
8. Collaborate with orthopedic specialists to monitor for age specific musculoskeletal problems. (Orthopedic Guidelines)

Mobility Guidelines

1. Develop an orthopedic transition plan. (clinical consensus) (Mobility Guidelines).
2. Counsel the patient about potential orthopedic degenerative problems. Consider bracing across the knee, such as the use of a KAFO, for patients with coronal plane valgus knee stress, or adding forearm crutches to decrease coronal and transverse plane trunk motion.
3. Counsel the patient about fractures and related precautions. (clinical consensus)

Physical Activity Guidelines

1. Discuss the National Physical Activity Guidelines with adults with Spina Bifida. (Health Promotion and Preventive Health Care Services Guidelines)
2. Follow the guidelines for adults with Spina Bifida as closely as possible, unless a health care provider advises that they are medically unsafe. (clinical consensus)
3. For substantial health benefits, it is recommended that adults should do at least 150 minutes (2 hours and 30 minutes) a week of moderate-intensity, or 75 minutes (1 hour and 15 minutes) a week of vigorous-intensity aerobic physical activity, or an equivalent combination of moderate- and vigorous-intensity aerobic activity. It is recommended that aerobic activity should be

performed in episodes of at least 10 minutes, and preferably, it should be spread throughout the week. (clinical consensus)

4. For additional and more extensive health benefits, it is recommended that adults should increase their physical activity to 300 minutes (5 hours) a week of moderate-intensity aerobic exercise, or 150 minutes (2 hours and 30 minutes) a week of vigorous-intensity aerobic physical activity, or an equivalent combination of moderate- and vigorous-intensity activity. Additional health benefits are gained by engaging in physical activity beyond this amount. (clinical consensus)

5. It is recommended that adults should include muscle-strengthening activities that involve all major muscle groups on 2 or more days a week. (clinical consensus)

6. It is recommended that all adults should avoid inactivity. Some physical activity is better than none, and adults who participate in any amount of physical activity gain some health benefits. (clinical consensus)

7. Identify and provide additional support and information on physical activity precautions for adults with shunts and ambulatory limitations.

8. Use health care encounters and follow-up meetings to develop physical activity goals and monitor progress (e.g. minutes of physical activity per day). Employ motivational interviewing techniques to discuss and set physical activity goals and strategies to overcome barriers to achieving those goals. Discuss the importance of physical activity and physical activity options with adults with Spina Bifida.

9. Prescribe, using a prescription pad, physical activity based off on goals discussed with adults with Spina Bifida.

10. Assist students who are considering post-secondary education to assess supports for physical activity in the educational institutions they are considering.

11. Use a team approach and include PTs/OTs to work with the adult with Spina Bifida to make sure that their mobility equipment fits properly in order to maximize their participation in physical activities. (clinical consensus)

12. Emphasize that any movement is beneficial.

Men's Health Guidelines

1. Conduct annual scrotal exam that documents testicular position, size, consistency, symmetry, and presence/absence of masses.

2. Assess and document genital sensation (penile, scrotal) yearly

3. Instruct patients on monthly testicular self-examination (TSE) to age 40 (clinical consensus)

4. Inform patients that sexual function and reproductive capacity may be altered as a sequela of Spina Bifida.(clinical consensus)

5. Provide information about safe sexual practices and genetic risk factors. (clinical consensus) (Sexual Health and Education Guidelines)

6. Refer the man to a urologist with expertise in male sexual function if he expresses concern regarding sexual dysfunction or an exam suggests impaired sensation or function of the genitalia. (clinical consensus) Similarly, it is recommended to make an appropriate referral to a specialist in male sexual function and/ or male infertility if he expresses concern related to orgasmic or ejaculatory function. (clinical consensus)

7. Characterize and record erectile function, orgasmic and ejaculatory function when relevant. (clinical consensus)
8. Explain to men with Spina Bifida that phosphodiesterase inhibitors are first-line pharmacologic treatments for erectile dysfunction. Men should be offered these treatments and instructed on their use if they do not have contraindications. (clinical consensus)
9. Employ open-ended questions to explore the man's interest in paternity and concerns about the heritability of Spina Bifida. Offer genetic counseling and infertility evaluation when questions about these topics arise. (clinical consensus)
10. Educate men about the risk of heritability of Spina Bifida for their offspring and offer their female partners additional supplementation with folic acid to reduce the risk. (clinical consensus) (Women's Health Guidelines)
11. Do not offer PSA testing to men with a life expectancy of less than 10-15 years or to men who are <55 and >69 years of age unless they are at elevated risk for prostate cancer based on family history.
12. For men between the ages of 55-69 with neuropathic bladder and chronic bacteriuria with at least 10-15 years life expectancy, the value of PSA alone as a screening tool is low. Discuss and offer PSA and digital rectal exam (DRE) testing as appropriate.
13. If a biopsy is recommended, consider using MRI-guidance, transperineal technique, and pre-treat men with culture-specific antibiotics prior to biopsy.
14. Consider waiting for fPSA normalization and tPSA nadir, typically about 12 weeks, before determining whether a biopsy should be performed based on elevated PSA in men with congenital neuropathic bladder on ISC who had a recent urinary tract infection.
15. Adequately assess pre-treatment bowel, urinary, and sexual function to guide counseling about treatment options for prostate cancer. (clinical consensus)
16. Prior to decision-making for treatment of prostate cancer, men with Spina Bifida may benefit from adjunct testing to fully characterize the risks of various treatments (e.g., cystourethroscopy to evaluate the external sphincter or urodynamics to evaluate bladder storage function). (clinical consensus)

Sexual Health and Education

1. Acknowledge that sexual health is an important part of adult life.
2. Take a history of sexual interest, functioning, experience and problems. (Men's Health Guidelines, Women's Health Guidelines – specifically information about fertility, reproduction, and anatomic functioning.)
3. Use factual information to educate adults about sexual health including intimate partner violence and sexual assault. (clinical consensus)
4. Provide guidance about safe sex practices including 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 physician or family medicine practitioner. Refer men to a sexual function clinic if desired. (clinical consensus) (Men's Health Guidelines, Women's Health Guidelines)
6. Educate about heritability of Spina Bifida. (Men's Health Guidelines, Women's Health Guidelines)

7. Create an environment in which the adult feels comfortable and safe discussing sexuality and sexual health routinely and openly during health care visits. (clinical consensus)
8. Refer to support groups and general audience literature regarding disability and sexuality. (clinical consensus)
9. Provide visual samples of items to facilitate discussions, such as female and male condoms, relevant websites, and other online resources. (clinical consensus)
10. Educate about the role of self-examination and routine health maintenance visits. (clinical consensus) (Men's Health Guidelines, Women's Health Guidelines)
11. Underscore goal of continence (Bowel Function and Care Guidelines, Urology Guidelines) for optimal sexual relationships. (clinical consensus)

Urology Guidelines

1. Obtain a renal/bladder ultrasound yearly. (clinical consensus)
2. Obtain a renal/bladder ultrasound, as needed if the adult has recurring symptomatic UTIs or if urodynamic testing identifies bladder hostility. (clinical consensus)
3. Obtain a serum creatinine test yearly. If the adult has low muscle mass, consider an alternative measure of renal function. (clinical consensus) (Self-Management and Independence Guidelines)
4. Obtain serum chemistries including B12 on anyone who has had urinary reconstruction. (clinical consensus)
5. Undertake cystoscopy and appropriate upper tract imaging in adults who have had a bladder augmentation when the following are present: (clinical consensus)
 - clinically-noted change in upper or lower urinary tract status
 - gross hematuria
 - recurrent symptomatic UTIs
 - increasing incontinence
 - pelvic pain
 - the adult has had a renal transplant with the presence of BK/polyomavirus
6. Evaluate patterns of continence/incontinence and address issues collaboratively with the individual and family. Include assessment of amount (volume) of incontinence as the amount in adults may be more bothersome than frequency.
7. Continue to support self-management and independent living. (Self-Management and Independence 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. Encourage women with Spina Bifida to have routine gynecological care, including Pap smears and mammograms.
3. Provide guidance on sexual health, education and birth control. (Sexual Health and Education Guidelines) Contraception options should be made available and discussed 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. Raise awareness of availability of screening and treatment for sexually transmitted infections, and offered Pap smears as per guidelines.

5. Provide gynecology exam rooms and tables that are accessible for women with physical disabilities. (clinical consensus)

6. Clinicians should initiate a discussion of sexual function with women with Spina Bifida in a sensitive manner to facilitate problem problem-solving and acknowledge common concerns such as inability to orgasm, prolonged time to achieve an orgasm, and decreased lubrication. In some cases, an experienced sexologist may be helpful.

7. Encourage women with decreased pelvic sensation to explore other parts of the body with themselves or a partner, especially the lips, nipples, arms, and earlobes, or other areas of the skin, as they may find them to be more sensitive.

8. Inform women that they can use commercially available sexual lubricants to improve lubrication.

9. Women with urinary incontinence should be encouraged to catheterize or void before having sex to prevent incontinence during sex.

10. Inform women that their choice of sexual positions may need to take into account their level of mobility, osteoporosis and fracture risk, and respiratory function.

11. Motivate women with Spina Bifida to communicate with their sexual partners about what they enjoy and do not enjoy during sex.

12. Recommend preconception consultation with an obstetrician who specializes in high-risk pregnancies. Depending on the woman's medical history, she may also benefit from preconception consultation with her neurosurgeon, urologist, physiatrists, and other health care providers to discuss the potential impact of pregnancy on health. (clinical consensus) (Prenatal Counseling Guidelines)

13. Recommend that women with Spina Bifida, who are at increased risk of having a baby with a neural tube defect, decrease their risk by taking a daily oral supplement of 4mg of folic acid starting at least 1 month but preferably 3 months prior to conception and continuing until 12 weeks of gestational age. (Prenatal Counseling Guidelines)

Pregnancy

1. Discuss the increased risk of preterm birth and review the signs and symptoms of preterm birth in the context of the woman's sensory abilities.

2. Inform women that maternal Spina Bifida increases a woman's chance of having a baby with Spina Bifida. (Prenatal Counseling Guidelines)

3. Conduct pulmonary function testing at least once during pregnancy in the case of kyphoscoliosis. This is because dyspnea can occur during pregnancy when there is an associated kyphoscoliosis deformity. (clinical consensus)

4. Ask about symptoms of shortness of breath at each antenatal visit, and undertake pulmonary function testing or assess for pulmonary embolism as indicated.

5. Consider temporary wheelchair use in women and girls who use braces and crutches to ambulate, to reduce the risk of falls and subsequently trauma to maternal joints and the fetus.

6. Follow for back and leg pain and consider temporary wheelchair use (clinical consensus), modified bedrest, and massage and physical therapy if back and leg pain is severe.

7. Consider referral to orthopaedics and physical medicine and rehabilitation as needed when there are significant or concerning changes in mobility. (clinical consensus)

8. Consider referral to occupational therapy and physical therapy early in pregnancy to discuss the impact of pregnancy on self-management ability as well as to discuss plans for after-delivery care and care of her baby. (Self-Management and Independence Guidelines)

9. Discuss bowel care early in the pregnancy, as pregnancy can worsen constipation. A diet high in fibre, increased fluid intake and exercise can alleviate constipation, however are not always sufficient. Bulk-forming agents such as psyllium, stool softeners such as docusate sodium, lubricant laxatives, osmotic laxatives and stimulate laxatives are considered safe in pregnancy. However, osmotic and stimulant laxatives may cause significant abdominal cramping and bloating and therefore should not be used for a prolonged period of time.

10. Consider having a consultation with a gastroenterologist or expert in neurogenic bowel management to maximize the methods to alleviate constipation. (clinical consensus)

11. Manage a suspected bowel obstruction with a team consisting of a general surgeon, neurosurgeon, and high-risk obstetrician. (clinical consensus)

12. Review signs of increased pressure, headache, nausea, and vomiting at each prenatal visit because the enlarging uterus can cause a shunt malfunction by increasing intra-abdominal pressure.

13. Manage signs of shunt malfunction with a team consisting of a neurosurgeon, obstetrician and anesthesiologist. Other specialties may be needed depending on the clinical scenario.

14. Conduct a thorough workup for both preeclampsia and shunt obstruction if a pregnant woman with a shunt has nausea, vomiting, headache, or neurological symptoms. A preeclampsia workup consists of assessing for the following: fetal well-being; blood pressure; proteinuria; and blood work to test for elevated aspartate aminotransferase (AST), and alanine transaminase (ALT), and thrombocytopenia.

15. Optimize medical management of seizures prior to conception. Women who have a history of seizures have a higher risk of seizure during pregnancy and labor. If possible, avoid anticonvulsant medications that have a greater risk of teratogenicity while still providing good control.

16. Perform regular urinalysis and urine culture tests throughout the pregnancy and treat infections promptly, as urinary tract infections are common during pregnancy in mothers with Spina Bifida.

17. Make a baseline renal assessment, ideally prior to pregnancy or early in the pregnancy, in order to make appropriate referrals to nephrology care.

18. Coordinate with a nephrologist to manage women with Spina Bifida who already have evidence of renal disease and a risk of decreased renal function in pregnancy.

19. Perform intensified maternal and fetal monitoring with women who have renal disease in pregnancy and are at increased risk of preeclampsia and intrauterine growth restriction.

19. Ask women at each visit about their ability to catheterize, and refer them to a urologist if there are concerns because urostomies can develop poor conduit drainage as the uterus grows.

20. Urgently consult with urology specialists if women with continent urinary diversions develop increased incontinence or difficulties in intermittent self-catheterization.

Childbirth

1. Consult a high-risk obstetrician when planning the mode of delivery. Although vaginal births are possible for women with Spina Bifida, severe spinal and pelvic skeletal deformities may prevent vaginal birth.

2. Consider facilitating vaginal deliveries in women with ventriculoperitoneal (VP) shunts by means of a shortened pushing stage, possibly aided by a vacuum or forceps to decrease elevation of intracranial pressure.

3. Teach women who may be unaware of labor contractions to palpate for hardening of the belly and observe for rupture of membranes, and watch for signs of autonomic dysreflexia.

4. Watch for autonomic dysreflexia triggered by labor among women who have a lesion above T6. Autonomic dysreflexia can be life-threatening and women experiencing any signs or symptoms should seek emergency care and transportation to the hospital. As well, there is significant clinical overlap between autonomic dysreflexia and preeclampsia, and therefore the woman should be evaluated for both.

5. Make the decision between a planned caesarean birth (with available urology back-up if needed) versus a planned trial of a vaginal birth (with the associated risks of having an emergency caesarean birth) in conjunction with a team consisting of an anesthesiologist, urologist, and obstetrician, and acknowledge the woman's goals and preferences. Keep in mind that a caesarean birth in a woman with previous lower urinary tract surgery may be complex. Intestinal and omental adhesions to the lower uterine segment may necessitate a classic upper segment section.

6. Recommend a caesarean birth to protect continence for women with vesical neck reconstruction or artificial sphincter placement.

7. Take into account that pregnancy itself can exacerbate an existing pelvic organ prolapse and that a vaginal birth will likely exacerbate it. Consider the impact of a worsening pelvic organ prolapse, and the possible need for subsequent surgery, in consultation with an obstetrician and urogynecologist, and taking into account the woman's preferences. The plan for the mode of birth should take into account the impact of this worsening and the possible need for subsequent surgery in consultation with an obstetrician and urogynecologist, and acknowledge the woman's preferences.

8. Consider that Spina Bifida can be associated with congenital renal malformations such as horseshoe kidney and pelvic kidney. If a caesarean birth is required, the surgeon should be aware of unique renal anatomy prior to conducting the surgery if needed.

9. Ensure that a consultant urologist be available for the caesarean birth in women who have had a previous lower urinary tract surgery.

10. Keep in mind that Spina Bifida is not a contraindication to epidural anesthesia. As such, ensure that each woman has an anesthesia consultation prior to delivery to discuss the risks and benefits of regional versus general anesthesia.

Breastfeeding

1. Encourage mothers who wish to breastfeed to do so and provide them with support from a lactation consultant. Keep in mind that there is no literature specifically about breastfeeding in the context of Spina Bifida. (clinical consensus) (Nutrition, Metabolic Syndrome, and Obesity Guidelines)

2. Be aware that while anti-epileptic medications are for the most part considered compatible with breastfeeding, some require close monitoring of the baby for side effects and a reduction in the baby's exposure. Consider informing mothers of any possible side effects associated with an anti-epileptic medication they are taking while breastfeeding.

Menopause

1. Inform women that vasomotor symptoms such as hot flashes can sometimes be managed by lifestyle changes such as avoiding alcohol, cigarette smoking and warm drinks, as well as maintaining a normal body mass index.

2. Take into account that medical management of vasomotor symptoms includes both hormonal and non-hormonal prescription medication. Decisions on which medication to take should be made in conjunction with a physician experienced in managing menopausal symptoms, and take into consideration the severity of the woman's symptoms, bone mineral density, risk for blood clots, and behavioral or emotional symptoms such as depression.

3. Inform women with vaginal dryness that they may benefit from topical vaginal lubricants.

4. Consider treating vaginal atrophy with vaginal estrogen by a physician experienced in managing the symptoms of menopause. This may also help with urinary urge incontinence and may prevent some urinary tract infections.

5. Women should be made aware of their breast anatomy, and should be encouraged to bring any changes to the attention of their physician.

6. Women should participate in breast cancer screening programs, which for many women will begin at age 45. This may be initiated sooner if there is a family history or other risk factors for breast cancers.

7. Women should continue to participate in cervical cancer screening programs in accordance with local guidelines.

8. Women should be made aware that abnormal vaginal perimenopausal bleeding and post-menopausal bleeding can be a sign of endometrial cancer. Strongly encourage women to tell their physician if they experience abnormal perimenopausal bleeding or any spotting or bleeding after menopause.

Deep Vein Thrombosis

1. Consider thromboprophylaxis on a case-by-case basis for girls and women with limited mobility and those who use wheelchairs. Girls and women with decreased mobility may have an increased risk of deep vein thrombosis and pulmonary embolism in pregnancy. Girls and women with thrombophilia; BMI>30; those who smoke; those with pelvic girdle pain that restricts mobility; those undergoing caesarean section or prolonged labor; those with preeclampsia; and those with a preterm birth are at further risk. Consider consultation with hematology to assist with risk assessment and thromboprophylaxis duration.

Bowel Function and Care Guidelines

1. Discuss consequences of constipation and bowel incontinence (including shunt malfunction, urinary incontinence, UTIs, skin breakdown, social isolation).
2. Establish the goal of bowel continence and institute the bowel continence program using timing, suppositories, pharmacologic agents or enemas as needed.
3. Assist the adult 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. Discuss management of bowel program as it may impact sexual relations.
7. Focus on fiber, fluids, exercise, and timed bowel movements after meals.
8. Consider a twofold attack of oral and rectal interventions to meet goal of bowel continence without constipation or fecal incontinence.
9. Use dietary management (fiber, fiber supplements, and fluids), pharmacologic adjuncts (sennoside, polyethylene glycol, lubiprostone, or other prescription), and/or rectal stimulants (glycerin, docusate sodium, or bisacodyl suppositories) to manage constipation.
10. Discuss other options for treatment if the above have failed, including cone enema or other transanal irrigation, cecostomy, or MACE.
11. Refer to a Spina Bifida clinic or specialist with expertise in bowel management in Spina Bifida. (clinical consensus)
12. Access support services for personal care if needed.

Endocrine: Puberty and Precocious Puberty Guidelines

1. 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 adult is more comfortable with a same-sex provider. (clinical consensus)
2. Document all positive and negative findings of the physical exam. (clinical consensus)
3. Discuss the outcomes of the evaluation with the patient and also with parents or caregivers, if appropriate, asking them if they have any concerns. (clinical consensus)
4. Consider a referral to a mental health professional if the individual is having psychosocial issues with their growth or development. (clinical consensus)
5. Discuss sexual health issues and make appropriate referrals to urologists, gynecologists or other sub-specialists such as endocrinology, adolescent medicine, genetics or others, as clinically appropriate. (clinical consensus) (Men's Health Guidelines, Sexual Health and Education Guidelines, Women's Health Guidelines)

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. Urge adults with Spina Bifida to continue following latex precautions, even if they have not experienced an adverse response to latex products (for example, latex-free condoms), until better scientific explanations are available to specifically drive prevention and intervention. (Sexual Health and Education Guidelines)
2. Educate adults 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)
3. Discuss avoidance of natural rubber products in the home and work environments.
4. Adults identified as having a latex allergy should have diphenhydramine and self-administered epinephrine available at all times. (clinical consensus)
5. Instruct adults to check food preparation in public venues as it should be prepared with latex-free gloves. (clinical consensus)
6. Educate adults about latex safe contraceptive products before they decide to become sexually active. (clinical consensus) (Sexual Health and Education Guidelines) (Appendix 1)
7. Review principles of latex precaution with the adult during a clinic visit and answer any questions. (clinical consensus)
8. If a person that is allergic to latex 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. Many of the positive tests may be due to laboratory cross-reactivity, but a clinical response of allergy will not be provoked. (Appendix 2)

Nutrition, Metabolic Syndrome, and Obesity Guidelines

1. Conduct annual assessments of weight, height or arm span, and calculate BMI. (clinical consensus) (Appendix: BMI and Body Composition Measurements)
2. However, explain that BMI is not accurate for people with paralysis, who have lowered ratios of fat to lean muscle tissue and that looking at the trajectory over time may be more useful.
3. Consider monitoring other measures of adiposity, such as waist circumference. (Appendix: BMI and Body Composition Measurements)
4. Conduct an annual assessment of blood pressure or blood pressure percentiles to monitor for pre-hypertension and hypertension. (clinical consensus)
5. Tailor the discussion around healthy nutrition to the adult's context. 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.
 - Identify who requires the information about healthy food (i.e. the adult with Spina Bifida, the caregiver, the attendant, the family member, or others).
 - Discuss the adult's existing access to cooking options and food preparation areas. (clinical consensus)
 - Involve a social worker or disability organization representative who can speak to adults about available local, state, and federal nutritional benefits such as the Supplemental Nutrition Assistance Program (SNAP), farmer's market vouchers or coupons, and other sorts of food vouchers that are available for eligible individuals.
 - Consider referral to a "Healthy Lifestyle" program and/or use a smartphone application, while recognizing that few such programs are tailored to individuals with disabilities (clinical consensus).
6. Provide information about potential interactions between nutrition in foods and medications.
 - Highlight that some medications, such as corticosteroids, have side-effects including weight gain, increased appetite, high blood pressure and a higher risk of developing osteoporosis or diabetes.
 - Provide information about specific foods and beverages that may interact with medications, such as antihypertensive, anticoagulant, or corticosteroid medications.
 - Encourage adults to disclose any prescribed, over-the-counter or complementary and alternative medications they are taking to all of their health care professionals, including pharmacists. (clinical consensus)
 - Emphasize the importance of reading medication labels to identify any dietary contraindications.¹ If this is difficult, discuss other ways that the adults could find out about potential contraindications, such as making the medication labels available in a larger font or asking the pharmacists for assistance.

7. Screening for abnormal blood glucose is indicated as part of assessing cardiovascular risk assessment in adults aged 40 to 70 years who have a BMI > 25 kg/m². Persons who have a family history of diabetes, have a history of gestational diabetes or polycystic ovarian syndrome, or are members of high risk racial/ethnic groups may be at increased risk for diabetes at a younger age or at a lower body mass index. Clinicians should consider screening earlier in persons with one or more of these characteristics.

8. Screening for dyslipidemia (fasting plasma profile) is recommended for men ≥ 40 years of age, and women ≥ 50 years of age or postmenopausal. Adults with the following risk factors should be screened at any age: current cigarette smoking, diabetes, arterial hypertension, family history of premature coronary heart disease, family history of hyperlipidemia, high risk ethnicity (individuals of First Nations or of South Asian ancestry), or the presence of rheumatoid arthritis, systemic lupus erythematosus, psoriatic arthritis, ankylosing spondylitis, inflammatory bowel disease, chronic obstructive pulmonary disease, chronic HIV infection, chronic kidney disease, abdominal aneurysm, or erectile dysfunction.

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 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.