

Guidelines for the Care of People with Spina Bifida

Health Promotion and Preventive Health Care Services

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Introduction

Individuals with Spina Bifida have unique and specific medical and social needs when compared to the general population, especially over a lifetime. They often have issues with pain, including the shoulders¹ and back, as well as neuropathic pain.² Neurogenic bowel and neurogenic bladder are common medical problems for people with Spina Bifida. Without routine monitoring, neurogenic bladders can result in kidney failure and require dialysis.³ Persons with Spina Bifida are also at increased risk for hypertension.^{4,5} Depending on the subtype of Spina Bifida there is also the risk of hydrocephalus, either with or without shunting, and this requires lifelong management.^{6,7} Self-management of personal medical care by adults with Spina Bifida may be difficult because of the frequent presence of cognitive impairment and the number and complexity of possible medical issues to be managed.⁸

Because of challenges with self-management and the need for environmental and attitudinal accessibility, individuals with Spina Bifida may find it difficult to access both routine and preventive health care services fully. Health professionals must be willing and able to take the time needed to appropriately communicate with and manage the sometimes complex health care needs of people with Spina Bifida.

Persons with Spina Bifida need the same routine screening procedures that any other person their age would need.⁹⁻¹⁰ However, many medical offices do not follow universal design principles. For example, examination rooms may be far from the reception area and/or otherwise inaccessible; examination tables may be too high or not be adjustable to allow for a transfer for a physical examination; scales may not be wheelchair accessible or safe for people with limited balance.

Persons with Spina Bifida also have difficulty with medical professionals' lack of knowledge about or recognition of common medical conditions related to Spina Bifida. Medical professionals who do not routinely care for people with Spina Bifida are less likely to recognize these unique situations and address them appropriately. Routine histories may not always include questions about possible changes in mobility or other bodily functions.

The goal of improving access for people with Spina Bifida is to promote better use of routine and preventive health. Health promotion supports improved long-term health and well-being, improved quality of life, and decreased utilization of emergency services and hospitalizations.^{7,11}

Outcomes

Primary

1. Maximize physical and mental health for individuals with Spina Bifida within the context of the underlying condition.
2. Identify risks and presence of common or known secondary conditions early.

Secondary

1. Limit preventable emergency department visits and hospitalizations for individuals with Spina Bifida.
2. Monitor trends of identified and newly-emerging secondary conditions.

Tertiary

1. Provide patient-centered general health monitoring based on the United States Preventive Services Task Force (USPSTF) recommendations and Spina Bifida- specific biomedical information.

0-11 months

Clinical Questions

1. Do children with Spina Bifida receive preventive health care recommended by the USPSTF?¹

Guidelines

1. Inform families about the importance of routine pediatric care, developmental surveillance and anticipatory guidance (e.g., immunizations and vision and hearing screens).⁹⁻¹⁰
2. Provide age-typical health promotion counseling (e.g. counseling for car seats or other motor vehicle occupant restraints, water safety, and nutrition).⁹⁻¹⁰ Counseling should be individualized to accommodate for Spina Bifida comorbidities such as having a shunt, mobility impairments, orthopedic deformities, developmental delays, and bowel and bladder management.^{10,12} (Bowel Function and Care Guidelines, Mobility Guidelines, Neuropsychology Guidelines, Neurosurgery Guidelines, Orthopedics Guidelines, Urology Guidelines)
3. Counsel families about possible future medical and social needs related to living with Spina Bifida. Needs might include latex allergies,^{5,13} chronic urinary issues,^{3,13} problems with shunts, and achieving an inclusive environment.^{2,13} (Latex Allergy Guidelines, Mobility Guidelines, Neurosurgery Guidelines, Urology Guidelines)
4. Monitor the child for neglect and/or abuse.⁹⁻¹⁰ (Family Functioning Guidelines)

1-2 years 11 months

Clinical Questions

1. Do children with Spina Bifida typically receive preventive health care recommended by the USPSTF?¹

Guidelines

1. Inform families about the importance of routine pediatric care, developmental surveillance and anticipatory guidance (e.g., immunizations, vision and hearing screens).⁹⁻¹⁰
2. Provide age-typical health promotion counseling (e.g., counseling for car seats or other motor vehicle occupant restraints, water safety, and nutrition).⁹⁻¹⁰ Counseling should be individualized to accommodate for Spina Bifida comorbidities such as having a shunt, bowel and bladder management, mobility impairments, orthopedic deformities and developmental delays.^{10,12} (Bowel Function and Care Guidelines, Mobility Guidelines,

Neuropsychology Guidelines, Neurosurgery Guidelines, Orthopedics Guidelines, Urology Guidelines)

3. Counsel families about possible future medical and social needs related to living with Spina Bifida. Needs might include latex allergies,¹² chronic urinary issues,^{3,12} problems with shunts, achieving an inclusive environment, overweight/obesity risk,⁹ importance of physical and recreational activity,¹ managing unexpected changes in function, keeping regular medical appointments, and pain.^{2,13} (Bowel Function and Care Guidelines, Latex Guidelines, Nutrition, Metabolic Syndrome, and Obesity Guidelines, Physical Activity Guidelines, Urology Guidelines)
4. Monitor the child for neglect and/or abuse.⁹⁻¹⁰ (Family Functioning Guidelines)

3-5 years 11 months

Clinical Questions

1. Do children with Spina Bifida typically receive preventive health care recommended by the USPSTF?¹

Guidelines

1. Inform families about the importance of routine pediatric care, developmental surveillance and anticipatory guidance (e.g., immunizations and vision and hearing screens).⁹⁻¹⁰
2. Promote age-typical health promotion counseling (e.g., counseling for car seats or other motor vehicle occupant restraints, water safety, and skill development).⁹⁻¹⁰ Counseling should be individualized to accommodate for Spina Bifida comorbidities such as having a shunt, mobility impairments, orthopedic deformities, developmental delays, and bowel and bladder management.⁹⁻¹⁰ (Bowel Function and Care Guidelines, Mobility Guidelines, Neuropsychology Guidelines, Neurosurgery Guidelines, Orthopedics Guidelines, Urology Guidelines)
3. Counsel families about possible future medical and social needs related to living with Spina Bifida. Needs might include latex allergies⁵, chronic urinary issues,^{5,13} problems with shunts, achieving an inclusive environment, overweight/obesity risk,⁹ importance of physical and recreational activity,⁹ managing unexpected changes in function, keeping regular medical appointments, and pain.^{2,13} (Bowel Function and Care Guidelines, Latex Guidelines, Nutrition, Metabolic Syndrome, and Obesity Guidelines, Physical Activity Guidelines, Urology Guidelines)
4. Monitor the child for neglect and abuse.⁹⁻¹⁰ (Family Functioning Guidelines)

6-12 years 11 months

Clinical Questions

1. Do early discussions about maintaining health and using health promotion and prevention strategies facilitate later participation in those activities?
2. Do children with Spina Bifida typically receive preventive health care recommended by the USPSTF?⁹
3. When does pain become a common secondary condition? What are the characteristics that increase risk for pain complaints?
4. What are the characteristics that increase the risk for hypertension?

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)
 - Hypertension.^{4,10,14} 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)

- Iron deficiency.⁹⁻¹⁰
 - Lipid disorders.⁹⁻¹⁰
 - Overweight/obesity, including the role in hypertension.⁹ (Nutrition, Metabolic Syndrome, and Obesity Guidelines)
 - Abuse, neglect, and/or violence.¹⁰ (Family Functioning Guidelines)
 - Social isolation, anxiety, and depression.⁹⁻¹⁰ (Mental Health Guidelines) o Motor vehicle and wheelchair safety.^{4,9-10} (Mobility Guidelines)
2. Provide counseling about tobacco and illicit drug use and refer the family to an appropriate treatment program if needed.⁹⁻¹⁰
 3. Provide guidance on skin cancer prevention.⁹⁻¹⁰
 4. Provide information about adaptive physical and recreational activities keeping in mind the particular child's degree of mobility.⁷ (Physical Activity Guidelines)
 5. Provide information about accessible physical activity and recreational opportunities in the community.⁹ (Physical Activity Guidelines)
 6. Monitor for pain and changes in pain using an appropriate pain scale for the child's level of cognition and communication, as pain may not be clearly recognized due to the unique neurologic status of children with Spina Bifida. Be aware that depending on their cognitive status, the child may not be able to give specific answers to questions such as the severity, frequency and duration of the pain. Proceed with appropriate evaluation and treatment.^{3,9,16}
 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)
 - Shunt concerns.⁵ Ask about any neurologic changes.
 - Sleep apnea. Ask if sleeping is restful and if there are snoring or apneic pauses during sleep.⁹ (Sleep Related Breathing Disorders Guidelines)
 - Skeletal and limb deformity. Check for new issues with bracing, positioning, or function. (clinical consensus)
 - 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.³ (Bowel Function and Care Guidelines, Urology Guidelines)
 - Skin breakdown and pressure injury.⁷ 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)
 - Adaptive equipment needs, including for orthoses, crutches, walkers, and wheelchairs.^{3,6-7,9} Make referrals to necessary subspecialists. (clinical consensus) (Mobility Guidelines)
 - 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.⁷ (Care Coordination Guidelines)
 9. Educate families on early signs of chronic conditions related to Spina Bifida.^{5,7}

13-17 years 11 months

Clinical Questions

1. Do early discussions about maintaining health and using health promotion and prevention strategies facilitate later participation in those activities?
2. Do children with Spina Bifida typically receive preventive health care recommended by the USPSTF?⁹
3. What are the parameters to begin screening for metabolic syndrome?
4. What are parameters and modifications are needed (if any) to begin education related to sexually transmitted infections, partner violence, and human papillomavirus (HPV) immunization?
5. What characteristics may put a child in this age range at risk for low self-rated health and health-related quality of life (HRQOL)? Do increasing acute medical conditions affect this?⁵ Are there interventions or supports that may mitigate this?
6. When does sleep apnea become notable and what are the risks?⁷
7. Do interventions make a difference in weight control, participation in physical or recreational activities, and pain control?
8. Can emergency department or hospital admissions be avoided?^{3,9,6}
9. Do hypertension interventions make a difference?

Guidelines

1. Monitor that the child is making routine well-child visits to his or her primary care provider to receive age-appropriate health promotion and preventive services, including age-appropriate screenings for:
 - Hypertension.^{4,9-10} Since there is no agreement on blood pressure targets for patients with Spina Bifida, it is recommended that baseline blood pressure is monitored to know what is considered hypertensive for the particular child. (clinical consensus)
 - Iron deficiency.⁹⁻¹⁰
 - Lipid disorders.⁹⁻¹⁰
 - Overweight/obesity, including the role in hypertension.⁹⁻¹⁰ (Nutrition, Metabolic Syndrome, and Obesity Guidelines)
 - Abuse, neglect, and/or violence.¹⁰ (Family Functioning Guidelines)
 - Social Isolation, Anxiety, Depression.⁹⁻¹⁰ (Mental Health Guidelines)
 - Motor vehicle and wheelchair safety.^{4,9-10} (Mobility Guidelines)
 - Contraceptive use, pregnancy, and sexually transmitted diseases. (as age appropriate) ¹ (Sexual Health and Education Guidelines, Women's Health Guidelines, Men's Healthcare Guidelines)
2. Provide counseling about tobacco and illicit drug use and refer the family to an appropriate treatment program if needed.⁹⁻¹⁰
3. Provide guidance on skin cancer prevention.⁹⁻¹⁰
4. Provide information about adaptive physical and recreational activities keeping in mind the particular child's degree of mobility.⁷ (Physical Activity Guidelines)
5. Provide information about accessible physical activity and recreational opportunities in the community.⁹ (Physical Activity Guidelines)
6. Monitor for pain and changes in pain using an appropriate pain scale for the child's level of cognition and communication, as pain may not be clearly recognized due to the unique neurologic status of children with Spina Bifida. Be aware that depending on their cognitive status, the child may not be able to give specific answers to questions such as the severity, frequency and duration of the pain. Proceed with appropriate evaluation and treatment.^{2-3,6}

7. Monitor for comorbid conditions that are specific to children with Spina Bifida, both during visits that are specifically intended to monitor Spina Bifida conditions as well during well-child visits. (Neurosurgery Guidelines, Orthopedics Guidelines, Bowel Function and Care Guidelines, Urology Guidelines, Mobility Guidelines, Skin Care Guidelines)
 - Shunt concerns.⁴ Ask about any neurologic changes.
 - Sleep apnea. Ask if sleeping is restful and if there are snoring or apneic pauses during sleep.⁹
 - Skeletal and limb deformity. Check for new issues with bracing, positioning, or function. (clinical consensus)
 - Constipation, urinary tract infections (UTIs), renal function, and problems with bowel and bladder regimens. Provide prescriptions for routine bowel medications, treatment of recurring UTIs, monitor for adherence to bowel and bladder management program and changes in bowel/bladder function.³ (Bowel Function and Care Guidelines, Urology Guidelines)
 - Skin breakdown and pressure injury.⁷ 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)
 - Adaptive equipment needs, including for orthoses, crutches, walkers, and wheelchairs.^{3,6-7,9} Make referrals to necessary subspecialists. (clinical consensus) (Mobility Guidelines)
 - 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.⁷ (Care Coordination Guidelines)
9. Educate families on early signs of chronic conditions related to Spina Bifida.^{5,7}

18+ years

Clinical Questions

1. Do early discussions about maintaining health and using health promotion and prevention strategies facilitate later participation in those activities?
2. Do adults with Spina Bifida receive preventive health care recommended by the USPSTF?¹
3. What characteristics may put the adult with Spina Bifida at risk of low self-rated health and health-related quality of life? Do increasing acute medical conditions affect this?⁵ Are there interventions or supports that may mitigate this?
4. Can future health issues and health care utilization be predicted? What is the prevalence and risks for common and Spina Bifida-related conditions? What is the expected cost of care?
5. Can the number of emergency department or hospital admissions be mitigated?^{3,9,6}

Guidelines

1. Monitor that the adult is receiving typical and age-related health promotion and preventive services, including screening or counseling about:
 - Hypertension.^{4,9-10} 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.
 - Lipid disorders.⁹ Treat as needed.
 - Overweight/obesity. Counsel as to healthy diet and exercise habits.¹ (Nutrition, Metabolic Syndrome, and Obesity Guidelines)

- Cancer, including skin cancer.⁹ Promote age-appropriate screening.
 - Diabetes and metabolic syndrome.⁹ Screening and treatment as needed.
 - Fall prevention.⁹
 - Adaptive physical activity.⁷ (Physical Activity Guidelines)
 - Depression and anxiety.⁹ (Mental Health Guidelines)
 - Smoking and illicit drug use.¹
 - Hearing and vision.⁹
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:
- Shunt concerns.⁴ Monitor for neurologic changes.
 - Sleep apnea.⁹ May need a referral for pulmonary evaluation and sleep study.
 - Skeletal and limb deformity.⁷ Check for new problems with positioning or brace use and new pain.
 - Osteoporosis. Counsel about the need for weight-bearing activities.¹
 - Pain.^{2,15} Use age- and cognition-appropriate pain scale to assess.
 - Constipation, urinary tract infections (UTIs), renal function, and problems with bowel and bladder regimens.^{3,7} 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)
 - Skin breakdown and pressure injury.⁹ Encourage adults to conduct frequent skin checks and to shift their weight at least every two hours.
 - Lymphedema.¹⁶
 - Adaptive equipment needs such as for orthoses, crutches, walkers, and wheelchairs.^{2-3,7,9,16-19} Make referrals to necessary subspecialists.
3. Promote care coordination between Spina Bifida-specific subspecialists and primary care providers.⁷ (Care Coordination Guidelines)
4. Educate adults about early signs of chronic conditions related to Spina Bifida.^{5,7}
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.^{6-7,9} (Self- Management and Independence Guidelines)

Research Gaps

1. Can future health issues and health care utilization be predicted? What are the common health conditions that are preventable or easily amenable to interventions? What is prevalence and risks for common and Spina Bifida-related conditions? What are the

- common causes for preventable or unanticipated mortality? What is the expected cost of care?
2. What are the common physical and mental health conditions associated with emergency department visits and hospitalizations? What are key anticipatory guidance or management strategies to prevent the need for higher levels of care? Can emergency department or hospital admissions be avoided, and how? Does a medical home help to prevent admissions for all age groups?¹⁸
 3. What are the characteristics that may put an individual with Spina Bifida at risk of low self-rated health and HRQOL? Are there preventive care interventions or supports that may mitigate this?
 4. What adaptations to general care practice and the USPSTF recommendations do individuals with Spina Bifida need, taking into account patient-centered perspectives and biomedical information?
 5. What long-term care planning is needed to support the best health for adults with Spina Bifida?

References

1. Roehrig, S., & Like, G. (2008). Factors affecting shoulder pain in adolescents and young adults with spina bifida. *Pediatric Physical Therapy*, 20(3), 224-232.
2. Werhagen, L., Hultling, C., & Borg, K. (2010). Pain, especially neuropathic pain, in adults with spina bifida, and its relation to age, neurological level, completeness, gender and hydrocephalus. *Journal of rehabilitation medicine*, 42(4), 374-376.
3. Ouyang, L., Bolen, J., Valdez, R., Joseph, D., Baum, M. A., & Thibadeau, J. (2015). Characteristics and survival of patients with end stage renal disease and spina bifida in the United States Renal Data System. *The Journal of urology*, 193(2), 558-564.
4. Mazur, L., Lacy, B., & Wilsford, L. (2011). The prevalence of hypertension in children with spina bifida. *Acta Paediatrica*, 100(8), e80-e83.
5. Stepanczuk, B. C., Dicianno, B. E., & Webb, T. S. (2014). Young adults with spina bifida may have higher occurrence of prehypertension and hypertension. *American journal of physical medicine & rehabilitation*, 93(3), 200-206.
6. Piatt Jr, J. H. (2016). Adults with myelomeningocele and other forms of spinal dysraphism: hospital care in the United States since the turn of the millennium. *Journal of Neurosurgery: Spine*, 25(1), 69-77.
7. Dicianno, B. E., Kurowski, B. G., Yang, J. M. J., Chancellor, M. B., Bejjani, G. K., Fairman, A. D., ... & Sotirake, J. (2008). Rehabilitation and medical management of the adult with spina bifida. *American journal of physical medicine & rehabilitation*, 87(12), 1027-1050.
8. Sawin, K. J., Bellin, M. H., Roux, G., Buran, C. F., & Brei, T. J. (2009). The experience of self-management in adolescent women with spina bifida. *Rehabilitation Nursing*, 34(1), 26-38.
9. US Preventive Services Task Force. (2006). *The guide to clinical preventive services: recommendations of the US Preventive Services Tack Force*. Lippincott Williams & Wilkins.
10. Workgroup, B. F. P. S., & Committee on practice and ambulatory medicine. (2017). 2017 recommendations for preventive pediatric health care. *Pediatrics*, e20170254.
11. Webb, T. S. (2010). Optimizing health care for adults with spina bifida. *Developmental disabilities research reviews*, 16(1), 76-81.

12. Young, N. L., Sheridan, K., Burke, T. A., Mukherjee, S., & McCormick, A. (2013). Health outcomes among youths and adults with spina bifida. *The Journal of pediatrics*, 162(5), 993-998.
13. Oddson, B. E., Clancy, C. A., & McGrath, P. J. (2006). The role of pain in reduced quality of life and depressive symptomology in children with spina bifida. *The Clinical journal of pain*, 22(9), 784-789.
14. Mann, J. R., Royer, J. A., Turk, M. A., McDermott, S., Holland, M. M., Ozturk, O. D., ... & Thibadeau, J. K. (2015). Inpatient and emergency room visits for adolescents and young adults with spina bifida living in South Carolina. *PM&R*, 7(5), 499-511.
15. Clancy, C. A., McGrath, P. J., & Oddson, B. E. (2005). Pain in children and adolescents with spina bifida. *Developmental Medicine and Child Neurology*, 47(1), 27-34.
16. Garcia, A. M., & Dicianno, B. E. (2011). The frequency of lymphedema in an adult spina bifida population. *American journal of physical medicine & rehabilitation*, 90(2), 89-96.
17. Gabrielsson, H., Traav, M. K., & Cronqvist, A. (2015). Reflections on health of young adults with spina bifida: the contradictory path towards well-being in daily life. *Open Journal of Nursing*, 5(04), 303.
18. Oakeshott, P., Hunt, G. M., Poulton, A., & Reid, F. (2010). Expectation of life and unexpected death in open spina bifida: a 40-year complete, non-selective, longitudinal cohort study. *Developmental Medicine & Child Neurology*, 52(8), 749-753.
19. Young, N. L., Anselmo, L. A., Burke, T. A., McCormick, A., & Mukherjee, S. (2014). Youth and young adults with spina bifida: their utilization of physician and hospital services. *Archives of physical medicine and rehabilitation*, 95(3), 466-471.