

Spina Bifida Guideline

Health promotion and preventive health care service guidelines for the care of people with spina bifida

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Abstract. Individuals with Spina Bifida (SB) have unique lifelong medical and social needs. Thus, when considering how to promote health and offer preventive care, providers must adapt general healthcare screening and counseling recommendations to their patients' physical and cognitive impairments along with discerning how to monitor secondary or chronic conditions common to the population. This article provides an update on the health promotion and preventive health care guidelines developed as part of the Spina Bifida Association's fourth edition of the Guidelines for the Care of People with Spina Bifida. The guidelines highlight accommodations needed to promote general preventive health, common secondary/chronic conditions such as obesity, metabolic syndrome, hypertension, musculoskeletal pain, and considerations for preventing acute care utilization for the SB population throughout the lifespan. Further research is needed to understand the effectiveness of preventive care interventions in promoting positive health outcomes and mitigating potentially preventable acute care utilization.

Keywords: Spina bifida, myelomeningocele, disabilities, health promotion, preventative health

1. Introduction

Individuals with Spina Bifida (SB) have unique lifelong medical and social needs. Thus, when considering how to promote health and offer preventive screenings and counseling in this population, providers must adapt general care guidelines to the specific needs of individuals with SB and monitor for and address common SB-related conditions.

Individuals with SB should have age-based routine screening and health promotion counseling such as height and weight monitoring, vision and hearing

screening, and wellness visits [1,2]. However, many medical offices may not have accessible facilities or equipment to conduct necessary screenings such as adjustable examination tables to allow for transfer, scales that are wheelchair accessible or safe for people with limited balance, and equipment and training to measure height for a patient who is not able to stand. Due to these facility limitations, individuals with SB may have difficulty in fully accessing routine and preventive health care services.

Moreover, counseling patients with SB and their families may require additional time. Families of infants and children may have many questions about routine care. School age children and young adults may require additional counseling regarding education services to meet their learning and health accommodation needs.

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As more people with SB achieve adulthood, attention must be paid to typical reproductive health counseling and screening for cancer and other common adult conditions. Given that individuals with SB often have learning, executive function, or intellectual disability, adolescents and adults may need additional coaching and navigation support regarding medical care, community living, education and employment, and self-management [3,4].

To provide comprehensive preventive care for individuals with SB, both pediatric and adult care clinicians should be familiar with and be able to screen for, recognize, manage, and appropriately refer for SB-related conditions. However, general preventive care tools and templates likely will not include questions about SB-related conditions such as bladder and bowel management, pressure ulcers, shunt problems, or musculoskeletal pain and mobility changes. Thus, clinicians may need to develop additional tools and templates to regularly evaluate these concerns and plan for extra time to discuss care plans with patients, caregivers, and their other health providers. Additionally, it is important to maintain care coordination among subspecialists and primary care providers throughout the lifespan [5,6].

2. Guideline goals and outcomes

Aligning with the goals in the general population, the overall goal of health promotion and preventive care for individuals with SB is to support improved long-term health and optimize chronic conditions, promote well-being and quality of life, and decrease emergency department visits and hospitalizations particularly for preventable conditions [7,8]. To focus our guideline development and recommendations for the SB population, the workgroup devised the following primary, secondary, and tertiary goals:

Primary

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

Secondary

1. Limit potentially preventable emergency department visits and hospitalizations for individuals with SB.
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 SB-specific care needs.

3. Methods

These guidelines were developed as part of the SB Association's fourth edition of the Guidelines for the Care of People with SB [9]. The scientific methodology for guideline development is described by Dicianno, et al. [10]. In summary, workgroups were convened consisting of international, multidisciplinary teams of clinical and research experts to develop a framework for the guidelines by topics. Secondly, the executive committee conducted an extensive literature review, consisting of library search strategies and supplementation by articles contributed by workgroup members. For this topic, the following search strategy was used: myelomeningocele, lipomyelomeningocele, meningocele, neural tube defects, spinal dysraphism, or SB were combined with the terms obesity, nutrition, diet, therapeutic recreation, community participation, preventive, hypertension, lymphedema, pain, and health outcomes. Additionally, proxy terms for health outcomes such as emergency department visits and hospitalizations were included. General health guidelines from the United States Preventive Services Task Force (USPSTF) were also reviewed to determine adaptations needed to implement these recommendations for individuals with SB. The articles were grouped by methodology, being cognizant of the Oxford Grading of Recommendations Assessment, Development and Evaluation (GRADE) ratings for quality of evidence related to strength of recommendation [11]. When applicable, the evidence from the literature search was used to develop these guidelines, and where it was limited to inform SB-specific preventive care, general age-based health care guidelines were provided with additional expert clinical recommendations for SB.

4. Clinical questions

The workgroup reviewed the previous guidelines, noting the absence of health promotion as a topic. The goals and outcomes for this new topic were determined by the group, along with a list of pertinent clinical questions organized by age. These questions can be sep-

Table 1
Clinical questions that informed the health promotion and preventive health care service guidelines

| Age group | Clinical questions |
|-------------|--|
| 0–11 months | 1. Do children with SB receive preventive health care recommended by the USPSTF? |
| 1–2 years | 1. Do children with SB receive preventive health care recommended by the USPSTF? |
| 11 months | |
| 3–5 years | 1. Do children with SB receive preventive health care recommended by the USPSTF? |
| 11 months | |
| 6–12 years | 1. Do early discussions about maintaining health and using health promotion and prevention strategies facilitate later participation in those activities? |
| 11 months | 2. Do children with SB typically receive preventive health care recommended by the USPSTF? 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? |
| 13–17 years | 1. Do early discussions about maintaining health and using health promotion and prevention strategies facilitate later participation in those activities? |
| 11 months | 2. Do children with SB typically receive preventive health care recommended by the USPSTF? 3. What are parameters to begin screening for metabolic syndrome? 4. What 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? Are there interventions or supports that may mitigate this? 6. When does sleep apnea become notable and what are the risks? 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? 9. Do hypertension interventions make a difference? |
| 18+ years | 1. Do early discussions about maintaining health and using health promotion and prevention strategies facilitate later participation in those activities? 2. Do adults with SB receive preventive health care recommended by the USPSTF? 3. What characteristics may put the adult with SB at risk of low self-rated health and health-related quality of life? Do increasing acute medical conditions affect this? Are there interventions or supports that may mitigate this? 4. Can future health issues and health care utilization be predicted? What are the prevalence and risks for common and SB-related conditions? What is the expected cost of care? 5. Can the number of emergency department or hospital admissions be mitigated? |

arated into three categories: (1) whether individuals with SB receive general health promotion guidelines and how preventive counseling affects longitudinal outcomes, (2) age of onset of secondary and/or chronic health conditions and how screening/intervention affects outcomes, and (3) how to predict and mitigate acute care utilization. These questions guided the literature search and became the framework of the Health Promotion and Preventive Health Care Services guidelines Table 1 provides the guiding questions.

5. Results

Using the initial search terms, 19 articles were included for review, consisting of 13 observational studies (cohort or cross-sectional studies), 2 qualitative studies, 2 narrative literature reviews, and 2 health promotion guidelines. Evidence was limited, based on available publications and study design. Expert clinical opinion or reliance on general USPSTF recommendations were used for the majority of the recommendations. Conse-

quently, the guidelines for Health Promotion and Preventive Medicine are not above the “low” rating, according to GRADE determinations [11]. The recommended care guidelines for Health Promotion and Preventive Health Care Services for people with SB are listed in Table 2.

There were significant gaps in the literature addressing the rate at which individuals with SB received recommended preventive care and how preventive screening, counseling, and early intervention affected related outcomes. Qualitative studies reported the importance of fostering self-management, organizational skills, and a sense of independence among adolescents and young adults with SB [12,13]. However, there were no interventional studies found with this search strategy that targeted these themes or measured the rate at which preventive screening guidelines or reproductive health care guidelines were met. Having a medical home may increase access to preventive care and screening. However, a single Canadian Institute of Health Information data system study found less than a quarter of adults with SB participated in that system of care [14].

Table 2
Health promotion and preventive health care service guidelines

| Age group | Guidelines* | Evidence |
|-------------------------|--|---|
| 0–11 months | <ol style="list-style-type: none"> 1. Inform families about the importance of routine pediatric care, developmental surveillance, and anticipatory guidance [1,2]. 2. Provide age-typical health promotion counseling (e.g., counseling for car seats or other motor vehicle occupant restraints, water safety, and nutrition). Counseling should be individualized to accommodate for SB-related conditions such as having a shunt, bowel and bladder management, mobility impairments, orthopedic deformities and developmental delays [1,2]. 3. Counsel families about possible future medical and social needs related to living with SB. Needs might include latex allergies, chronic urinary issues, problems with shunts, skin ulcers, and overweight/obesity risk. Preventive counseling should include: strategies for physical and recreational activity, managing unexpected changes in function, and ways to achieve an inclusive environment at home and in the community [8,49,50]. 4. Monitor the child for neglect and/or abuse [1,2]. 5. Monitor for caregiver burnout. | Clinical consensus, references given where applicable |
| 1–2 years 11 months | <ol style="list-style-type: none"> 1. Inform families about the importance of routine pediatric care, developmental surveillance, and anticipatory guidance [1,2]. 2. Provide age-typical health promotion counseling (e.g., counseling for car seats or other motor vehicle occupant restraints, water safety, and nutrition). Counseling should be individualized to accommodate for SB-related conditions such as having a shunt, bowel and bladder management, mobility impairments, orthopedic deformities and developmental delays [1,2]. 3. Counsel families about possible future medical and social needs related to living with SB. Needs might include latex allergies, chronic urinary issues, problems with shunts, skin ulcers, and overweight/obesity risk. Preventive counseling should include: strategies for physical and recreational activity, managing unexpected changes in function, and ways to achieve an inclusive environment at home and in the community [8,49,50]. 4. Monitor the child for neglect and/or abuse [1,2]. 5. Monitor for caregiver burnout. | Clinical consensus, references given where applicable |
| 3–5 years 11 months | <ol style="list-style-type: none"> 1. Inform families about the importance of routine pediatric care, developmental surveillance, and anticipatory guidance [1,2]. 2. Provide age-typical health promotion counseling (e.g., counseling for car seats or other motor vehicle occupant restraints, water safety, and nutrition). Counseling should be individualized to accommodate for SB-related conditions such as having a shunt, bowel and bladder management, mobility impairments, orthopedic deformities and developmental delays [1,2]. 3. Counsel families about possible future medical and social needs related to living with SB. Needs might include latex allergies, chronic urinary issues, problems with shunts, skin ulcers, and overweight/obesity risk. Preventive counseling should include: strategies for physical and recreational activity, managing unexpected changes in function, and ways to achieve an inclusive environment at home and in the community [8,49,50]. 4. Monitor the child for neglect and/or abuse [1,2]. 5. Monitor for caregiver burnout. | Clinical consensus, references given where applicable |
| 6–12 years 11 months | <ol style="list-style-type: none"> 1. Review that the child is having routine well-child visits and age-appropriate health promotion and preventive services [2,51], including screenings for: <ul style="list-style-type: none"> – Hearing and Vision Screenings. Recognize that ophthalmic complications are common in SB including nystagmus, strabismus, optic atrophy, and papilledema. Vision and eye movement changes may be signs of shunt malfunction [52]. – Age-Based Immunizations – Dental Care – Hypertension [15,16] – Overweight/obesity. Use adjusted arm span or recumbent height measurement and wheelchair scale for those who are wheelchair dependent [53]. Screen for metabolic syndrome [17,33,54]. – Motor vehicle and wheelchair safety – may need adaptive car seat – Pressure ulcer and skin cancer prevention—encourage daily skin checks [55,56]. – Provide information about adaptive physical and recreational activities keeping in mind the child's degree of mobility. – Social isolation, Depression, Anxiety and need for counseling [57] – School and peer difficulties [58] – Reproductive health including early menarche for girls and sexual safety for teens [28,59–62]. – Abuse, neglect, and/or violence – Caregiver burnout | Clinical consensus, references given where applicable |

Table 2, continued

| Age group | Guidelines* | Evidence |
|-----------------------------|---|---|
| 13–17 years 11 months | <p>2. Monitor and provide preventive education for SB co-occurring conditions both during SB-specific and well-child visits:</p> <ul style="list-style-type: none"> – Neurologic changes which may indicate problems such as: shunt malfunction (headache, vision changes, poor concentration, vomiting), Chiari compression (neck pain/headache, breathing changes, swallowing changes, or apnea) or tethered cord (worsening scoliosis, pain, new weakness/numbness) [63,64]. – Sleep apnea. Ask if sleeping is restful and if there are snoring or apneic pauses during sleep [65]. – Bladder and bowel concerns: constipation, UTIs, renal function, and problems/adherence with bowel and bladder regimens [50,66,67]. – Skin breakdown and pressure injury – Encourage daily skin checks and routine weight transfers [56]. – Skeletal and limb deformity. Check for new issues with bracing, positioning, or function. – Adaptive equipment needs, including orthoses, crutches, walkers, wheelchairs, and lifts for transfers, toileting, and bathing. – 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 decreased sensation or cognitive ability [21,68]. <p>3. Promote care coordination among SB subspecialists and primary care providers [5,6].</p> | Clinical consensus, references given where applicable |
| | <p>1. Review that the child is having routine well-child visits and age-appropriate health promotion and preventive services [2,51], including screenings for:</p> <ul style="list-style-type: none"> – Hearing and Vision Screenings. Recognize that ophthalmic complications are common in SB including nystagmus, strabismus, optic atrophy, and papilledema. Vision and eye movement changes may be signs of shunt malfunction [52]. – Age-Based Immunizations. – Dental Care. – Hypertension [15,16]. – Overweight/obesity. Use adjusted arm span or recumbent height measurement and wheelchair scale for those who are wheelchair dependent (53). Screen for metabolic syndrome [17,33,54]. – Motor vehicle and wheelchair safety – may need adaptive car seat. – Pressure ulcer and skin cancer prevention – encourage daily skin checks [55,56]. – Provide information about adaptive physical and recreational activities keeping in mind the child’s degree of mobility. – Social isolation, Depression, Anxiety and need for counseling [57]. – School and peer difficulties [58]. – Reproductive health including early menarche for girls and sexual safety for teens [28,59–62]. – Abuse, neglect, and/or violence. – Caregiver burnout. <p>2. Monitor and provide preventive education for SB co-occurring conditions both during SB-specific and well-child visits:</p> <ul style="list-style-type: none"> – Neurologic changes which may indicate problems such as: shunt malfunction (headache, vision changes, poor concentration, vomiting), Chiari compression (neck pain/headache, breathing changes, swallowing changes, or apnea) or tethered cord (worsening scoliosis, pain, new weakness/numbness) [63,64]. – Sleep apnea. Ask if sleeping is restful and if there are snoring or apneic pauses during sleep [65]. – Bladder and bowel concerns: constipation, UTIs, renal function, and problems/adherence with bowel and bladder regimens [50,66,67]. – Skin breakdown and pressure injury – Encourage daily skin checks and routine weight transfers [56]. – Skeletal and limb deformity. Check for new issues with bracing, positioning, or function. – Adaptive equipment needs including orthoses, crutches, walkers, wheelchairs, and lifts for transfers, toileting, and bathing. – 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 decreased sensation or cognitive ability [21,68]. <p>3. Promote care coordination among SB subspecialists and primary care providers [5,6].</p> | |

Table 2, continued

| Age group | Guidelines* | Evidence |
|-----------|--|---|
| 18+ years | <ol style="list-style-type: none"> 1. Monitor that the adult patient is receiving typical and age-related health promotion and preventive services [2,51], including screening and counseling for: <ul style="list-style-type: none"> – Vision and Hearing Screenings. Recognize that ophthalmic complications are common in SB including nystagmus, strabismus, optic atrophy, and papilledema. Vision and eye movement changes may be signs of shunt malfunction [52]. – Age-based Immunizations. – Dental Care. – Hypertension [15,16]. – Overweight/obesity. Use adjusted arm span or recumbent height measurement and wheelchair scale for those who are wheelchair dependent [53]. – Screen for metabolic syndrome, lipid disorders, and diabetes [17,33,54]. – Cancer screening per UPTSF considering accommodations needed for physical or intellectual disability [29–31]. – Motor vehicle and wheelchair safety – may need adaptive car seat. – Fall prevention – safe transfers and mobility. – Adaptive physical activity. – Smoking and illicit drug use. – Reproductive health counseling [28,59–62]. Evaluate for sexual function concerns. Pelvic floor concerns such as prolapse. Provide counseling about family planning and possible fertility and genetic counseling to individuals interested in pregnancy. Recommend prenatal vitamins and 4 mg of folic acid prior to conception. – Depression and anxiety/need for counseling [69]. – Abuse, neglect, and/or violence. – Caregiver burnout. 2. Monitor and provide preventive education for SB co-occurring conditions both during chronic care follow up and annual preventive care visits including: <ul style="list-style-type: none"> – Neurologic changes which may indicate problems such as: shunt malfunction (headache, vision changes, poor concentration, vomiting), Chiari compression (neck pain/headache, breathing changes, swallowing changes, or apnea) or tethered cord (worsening scoliosis, pain, new weakness/numbness) [70]. – Sleep apnea – Evaluate for sleep disordered breathing, daytime sleepiness/poor concentration, elevated serum bicarbonate [65,71]. – Bladder and bowel concerns: constipation, UTIs, renal function, and problems/adherence with bowel and bladder regimens [50,66,67]. – Skeletal and limb deformity – Check for problems or pain with positioning or orthotic use. – Osteoporosis–Counsel about the risks, benefits, and limitations for weight-bearing activities [8,72]. – Pain – Use age and cognition-appropriate pain scale to assess. May also ask about pain interference in daily activities. – Skin breakdown and pressure injury – Encourage daily skin checks [56]. – Lymphedema – Consider compression socks or wound care referral [22]. – Adaptive equipment needs including orthoses, crutches, walkers, wheelchairs, and lifts for transfers, toileting, and bathing. 3. 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. 4. Promote care coordination among subspecialists and primary care providers [5,6]. | Clinical consensus, references given where applicable |

* Numbers in parentheses are articles in the Reference List. Of note, articles cited include those from the initial search strategy and additional ones found following the process.

Limited studies have evaluated the age of onset and the longitudinal outcomes for screening and early intervention of secondary and other chronic conditions such as hypertension, metabolic syndrome, pain, and sleep apnea in SB. While observational studies identified conditions such as hypertension [15,16] metabolic syndrome [17], pain [18–21], and lymphedema [22],

there were no studies evaluating interventions to decrease the risk or successfully treat conditions in the SB population.

Information from three secondary analyses of large databases note individuals with SB have increased emergency department and inpatient utilization compared to the general population [14,23,24]. Neurogenic

bladder related issues, especially urinary tract infections (UTI), may be the most common reasons for health care utilization [24,25]. They are implicated as a risk for mortality in individuals with SB who have end stage renal disease, are on dialysis, and are post transplantation [27]. A single study from the United Kingdom reported that death before age five was common, a higher level of SB lesion was associated with a higher risk for mortality. Later causes of death were most commonly documented as unknown, followed by epilepsy, pulmonary embolus, acute hydrocephalus, and urosepsis [26]. Of the articles found using the search criteria, none evaluated interventions to prevent emergency room use, inpatient admission, morbidity, or mortality.

6. Discussion

There is limited scientific information about health promotion and preventive health care services for people with SB. Although the majority of the guidelines regarding SB-specific care are based on clinical consensus, there is strong support for following the general USPSTF recommendations for wellness exams and screening in all age groups. Studies to determine if individuals with SB receive general recommended preventive care at the same frequency as those without SB are lacking. For children and adolescents with SB, regular follow up with a primary care or medical home clinic and a SB-specific multidisciplinary clinic may promote preventive care services and care coordination [25–27]. However, this care model may not be available for adults with SB [14]. Since the guideline search process was completed, there has been additional evidence of the limited preventive care for adults with SB related to reproductive health care and cancer screening [28–31]. Both men and women with SB may have poor understanding of their reproductive potential and face discouragement from their providers in discussing their reproductive wishes [28,32]. Thus, efforts to discuss reproductive health and safe sex practices with adolescents and adults with SB should be included in preventive care visits. Barriers to receiving age-appropriate screenings for adults with disability mirror those of the general population (e.g., lack of knowledge about screenings, lack of provider referral, time constraints, and poor experiences during screenings) [30]. Moreover, adults with disability may have unique barriers such as fearing the burden of a new diagnosis in addition to their underlying condition and lack of appropriate health care system accommodations

(e.g., transportation, equipment facilities, and personnel trained to aid them with transfers and positioning) [30]. Strategies to overcome such barriers are relevant for any provision of health care for people with disability and include training for professionals and their office staff regarding respectful communication, medical knowledge and skills about disability care, and proper equipment and facility accommodation [28,30]. These recommendations are for primary care as well as specialty care clinics.

Compared to the general population, individuals with SB may have increased common cardiovascular risks such as hypertension, metabolic syndrome, and obesity [15–17,33]. It should be noted that there is insufficient data to define the most accurate method for height measurement for individuals with SB, particularly for those with higher lesions who are wheelchair dependent. Use of other methods, such as waist circumference, may be a better option [34]. Consistency of measurement is important for counseling related to nutrition, physical activity and obesity management. For more information on nutrition and obesity as well as anthropomorphic measurements, please see the Nutrition, Metabolic Syndrome and Obesity Guidelines article in this special issue. Other common secondary/chronic conditions of concern for people with SB include pain, bowel/bladder management, pressure ulcers, depression, sleep disturbance, and limited social and community participation [35–37]. The most common medical condition reported in a US survey of adults with SB ($n = 852$) was pain within the past 7 days (46.9% of respondents), involving the low back and lower limbs [37]. Bowel and bladder concerns should also be addressed since they significantly impact quality of life and function [35,37,38] and are related to increased care utilization in all age groups [24,39,40]. Recurrent urinary tract infections occur in approximately 30% of individuals with SB and have implications for antibiotic stewardship and long-term renal function [37,41]. Large volume urinary incontinence impacts quality of life and sexual satisfaction for adults [36,42]. Whereas any fecal incontinence is associated with lower quality of life for all age groups and decreased employment [43,44]. Sleep apnea seems to be an emerging secondary/chronic condition for SB, although there are only a few studies. Rates of diagnosis of moderate to severe sleep apnea in two single site studies are reported as 3% of those tested (< 10% of patients who were tested) [30] and 30% [31], respectively. Thus, more studies are needed to determine the best screening strategies for sleep apnea. Evaluating for these conditions during child, adolescent, and

adult preventive care visits is recommended; however, further research is needed to assess the effectiveness of screening and intervention.

Individuals with SB experience increased morbidity and mortality as well as increased acute care utilization compared to those without SB [14,23,39,45,46]. Urinary tract infections, device complications such as shunt malfunction, pressure ulcers and soft tissue infections, as well as pneumonia are often reasons for acute care utilization for all age groups and therefore are important diagnoses to target for preventive care interventions [23,24,39,47]. However, intervention studies to decrease emergency and acute care use have been sparse. One wellness intervention utilizing home-based case management and goal-setting self-management initially demonstrated a decrease in all-cause emergency department use; however there was no significant change in emergency department use or hospital admissions after two years [48].

Although the scientific evidence is limited, clinical consensus recognizes the importance of identifying and evaluating SB-related conditions during prevention and chronic care visits (see Guidelines for the Care of People with SB on the www.spinabifidassociation.org website for more information). In acknowledgement of the limitations in the evidence regarding preventive health recommendations for the SB population, the workgroup developed the following questions to direct future research:

- What are the prevalence and the risks for common and SB-related conditions?
- What are the common causes for preventable or unanticipated mortality?
- What are key anticipatory guidance or management strategies to prevent the need for acute care utilization?
- How does a medical home help to prevent admissions for all age groups?
- What are the characteristics that may put an individual with SB at risk of low self-rated health and quality of life?
- Are there preventive care interventions or supports that may promote self-rated health or quality of life?
- What adaptations to general care practice and the USPSTF recommendations do individuals with SB need, taking into account patient-centered perspectives and biomedical information?
- What long-term care planning is needed to support the best health for adults with SB?

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The Spina Bifida Association has already embarked on a systematic process for reviewing and updating the guidelines. Future guidelines updates will be made available as they are completed.

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Conflict of interest

Margaret Turk, MD is Co-Editor-in-Chief for the *Disability and Health Journal*, and reports a cooperative agreement with the Centers for Disease Control and Prevention, National Center for Birth Defects and Developmental Disabilities (#1U19DD001218)The other authors do not have any conflicts of interest to disclose.

References

- [1] US Preventive Services Task Force. The guide to clinical preventive services: recommendations of the US Preventive Services Task Force. Lippincott Williams & Wilkin, 2006.
- [2] Committee on Practice and Ambulatory Medicine, Bright Futures Periodicity Schedule Workgroup. 2017 Recommendations for Preventive Pediatric Health Care. *Pediatrics*. 2017; 139(4): e20170254. doi: 10.1542/peds.2017-0254.
- [3] Hampton LE, Fletcher JM, Cirino PT, Blaser S, Kramer LA, Drake J, et al. Hydrocephalus status in spina bifida: an evaluation of variations in neuropsychological outcomes. *J Neurosurg Pediatr*. 2011 Sep; 8(3): 289-98. doi: 10.3171/2011.6.PEDS10584.
- [4] Burmeister R, Hannay HJ, Copeland K, Fletcher JM, Boudousquie A, Dennis M. Attention problems and executive functions in children with spina bifida and hydrocephalus. *Child Neuropsychol*. 2005; 11(3): 265-83. doi: 10.1080/092970490911324.
- [5] Brustrom J, Thibadeau J, John L, Liesmann J, Rose S. Care Coordination in the Spina Bifida Clinic Setting: Current Practice and Future Directions. *J Pediatr Health Care*. 2012; 26(1): 16-26. doi: 10.1016/j.pedhc.2010.06.003.
- [6] Thibadeau J, Walker WO, Castillo J, Dicianno BE, Routh JC, Smith KA, et al. Philosophy of care delivery for spina bifida. *Disabil Health J*. 2020; 13(2): 100883. doi: 10.1016/j.dhjo.2019.100883.
- [7] Dicianno BE, Kurowski BG, Yang JMJ, Chancellor MB, Bejjani GK, Fairman AD, et al. Rehabilitation and Medical Management of the Adult with Spina Bifida. *Am J Phys Med Rehabil*. 2008; 87(12): 1027-50. doi: 10.1097/PHM.0b013e31818de070.
- [8] Webb TS. Optimizing health care for adults with spina bifida. *Dev Disabil Res Rev*. 2010; 16(1): 76-81. doi: 10.1002/ddrr.99.
- [9] Spina Bifida Association. Guidelines for the Care of People with Spina Bifida [Internet]. 2018 Available from: <https://www.spinabifidaassociation.org/guidelines/pdf>.
- [10] Dicianno BE, Beierwaltes P, Dosa N, Raman L, Chelliah J, Struwe S, et al. Scientific methodology of the development of the Guidelines for the Care of People with Spina Bifida: An initiative of the Spina Bifida Association. *Disabil Health J*. 2020; 13(2): 100816. doi: 10.1016/j.dhjo.2019.06.005.
- [11] Guyatt GH, Oxman AD, Vist GE, Kunz R, Falck-Ytter Y, Alonso-Coello P, et al. GRADE: an emerging consensus on rating quality of evidence and strength of recommendations. *BMJ*. 2008; 336: 924-6. doi: 10.1136/bmj.39489.470347.AD.
- [12] Sawin KJ, Bellin MH, Roux G, Buran CF, Brei TJ. The Experience of Self-Management in Adolescent Women with Spina Bifida. *Rehabil Nurs*. 2009; 34: 26-38. doi: 10.1002/j.2048-7940.2009.tb00245.x.
- [13] Gabriellsson H, Traav MK, Cronqvist A. Reflections on Health of Young Adults with Spina Bifida: The Contradictory towards Well-being in Life. *Open Journal of Nursing*. 2015; 5(4): 303-312.
- [14] Young NL, Anselmo LA, Burke TA, McCormick A, Mukherjee S. Youth and young adults with spina bifida: Their utilization of physician and hospital services. *Arch Phys Med Rehabil*. 2014; 95(3): 466-71. doi: 10.1016/j.apmr.2013.09.015.
- [15] Stepanczuk BC, Dicianno BE, Webb TS. Young Adults with Spina Bifida May Have Higher Occurrence of Prehypertension and Hypertension. *Am J Phys Med Rehabil*. 2013; 92(11): 1-7. doi: 10.1097/PHM.0b013e3182a92b03.
- [16] Mazur L, Lacy B, Wilsford L. The prevalence of hypertension in children with spina bifida. *Acta Paediatr*. 2011; 100(8): e80-3. doi: 10.1111/j.1651-2227.2011.02225.x.
- [17] Van Speybroeck A, Mueske NM, Mittelman SD, Kremer RK, Ryan DD, Wren TAL. Fasting serum blood measures of bone and lipid metabolism in children with myelomeningocele for early detection of cardiovascular and bone fragility risk factors. *J Spinal Cord Med*. 2017; 40(2): 193-200. doi: 10.1080/10790268.2015.
- [18] Roehrig S, Like G. Factors affecting shoulder pain in adolescents and young adults with spina bifida. *Pediatr Phys Ther*. 2008; 20(3): 224-32. doi: 10.1097/PEP.0b013e318181162a.
- [19] Werhagen L, Hultling C, Borg K. Pain, especially neuropathic pain, in adults with spina bifida, and its relation to age, neurological level, completeness, gender and hydrocephalus. *J Rehabil Med*. 2010; 42(4): 374-6. doi: 10.2340/16501977-0529.
- [20] Clancy CA, McGrath PJ, Oddson BE. Pain in Children and Adolescents with Spina Bifida. *Dev Med Child Neurol*. 2005; 47: 27-34. doi: 10.1017/s0012162205000058.
- [21] Oddson BE, Clancy CA, McGrath PJ. The role of pain in reduced quality of life and depressive symptomatology in children with spina bifida. *Clin J Pain*. 2006; 16(2): 993-8. doi: 10.1097/01.ajp.0000210929.43192.5d.
- [22] Garcia AM, Dicianno BE. The frequency of lymphedema in an adult spina bifida population. *Am J Phys Med Rehabil*. 2011; 90(2): 89-96. doi: 10.1097/PHM.0b013e318201753e.
- [23] Mann JR, Royer JA, Turk MA, McDermott S, Holland MM, Ozturk OD, et al. Inpatient and Emergency Room Visits for Adolescents and Young Adults with Spina Bifida. *PM R*. 2015; 7: 499-511. doi: 10.1016/j.pmrj.2014.11.011.
- [24] Piatt JH. Adults with myelomeningocele and other forms of spinal dysraphism: hospital care in the United States since the turn of the millennium. *J Neurosurg Spine*. 2016; 25(1): 69-77. doi: 10.3171/2015.9.SPINE15771.
- [25] Burke R, Liptak GS, Council on Children with Disabilities. Providing a Primary Care Medical Home for Children and

- Youth With Spina Bifida. *Pediatrics*. 2011; 128(6): e1645-57. doi: 10.1542/peds.2011-2219.
- [26] Kaufman BA, Terbrock A, Winters N, Ito J, Klosterman A, Park TS. Disbanding a multidisciplinary clinic: effects on the health care of myelomeningocele patients. *Pediatr Neurosurg*. 1994; 21(1): 36-44. doi: 10.1159/000120812.
- [27] Akobirshoev I, Parish S, Mitra M, Dembo R. Impact of Medical Home on Health Care of Children With and Without Special Health Care Needs: Update from the 2016 National Survey of Children's Health. *Matern Child Health J*. 2019; 23(11): 1500-7. doi: 10.1007/s10995-019-02774-9.
- [28] Streur CS, Schafer CL, Garcia VP, Quint EH, Sandberg DE, Kalpakjian CZ, et al. "He told me it would be extremely selfish of me to even consider [having kids]": The importance of reproductive health to women with spina bifida and the lack of support from their providers. *Disabil Health J*. 2019; 13(2): 100815. doi: 10.1016/j.dhjo.2019.06.004.
- [29] Magasi S, Reis JP, Wilson T, Rosen A, Ferlin A, VanPuybrouck L. ScreenABLE: Breast Cancer Screening among Women with Disabilities from Community Identified Challenges to Community-Based Programs. *Prog Community Health Partnersh*. 2019; 13(5): 61-9. doi: 10.1353/cpr.2019.0039.
- [30] Edwards DJ, Sakellariou D, Anstey S. Barriers to, and facilitators of, access to cancer services and experiences of cancer care for adults with a physical disability: A mixed methods systematic review. *Disabil Health J*. 2020; 13(1): 100844. doi: 10.1016/j.dhjo.2019.100844.
- [31] Johnston AW, Wolf S, Alkazemi MH, Pomann GM, Wood H, Wiener JS, et al. Patterns of inpatient care for prostate cancer in men with spina bifida. *Disabil Health J*. 2020; 13(2): 100866. doi: 10.1016/j.dhjo.2019.100866.
- [32] Wiener JS, Frimberger DC, Wood H. Spina Bifida Health-care Guidelines for Men's Health. *Urology*. 2018; 116: 218-26. doi: 10.1016/j.urology.2018.01.005.
- [33] Nelson MD, Widman LM, Abresch RT, Stanhope K, Havel PJ, Styne DM, et al. Metabolic syndrome in adolescents with spinal cord dysfunction. *J Spinal Cord Med*. 2007; (30 Suppl 1)(7): S127-39. doi: 10.1080/10790268.2007.
- [34] Liu JS, Dong C, Vo AX, Dickmeyer LJ, Leung CL, Huang RA, et al. Obesity and anthropometry in spina bifida: What is the best measure. *J Spinal Cord Med*. 2018; 41(1): 55-62. doi: 10.1080/10790268.2016.1195071.
- [35] Wagner R, Linroth R, Gangl C, Mitchell N, Hall M, Cady R, et al. Perception of secondary conditions in adults with spina bifida and impact on daily life. *Disabil Health J*. 2015; 8(4): 492-8. doi: 10.1016/j.dhjo.2015.03.012.
- [36] Szymanski KM, Cain MP, Whittam B, Kaefer M, Rink RC, Misseri R. Incontinence affects health-related quality of life in children and adolescents with spina bifida. *J Pediatr Urol*. 2018; 14(3): 279.e1-279.e8. doi: 10.1016/j.jpuro.2018.02.021.
- [37] Morley CP, Struwe S, Pratte MA, Clayton GH, Wilson PE, Dicianno BE, et al. Survey of US adults with spina bifida. *Disabil Health J*. 2020; 13(2). doi: 10.1016/j.dhjo.2019.100833.
- [38] Szymanski KM, Cain MP, Whittam B, Kaefer M, Rink RC, Misseri R. Incontinence affects health-related quality of life in children and adolescents with spina bifida. *J Pediatr Urol*. 2018; 14(3): 279.e1-279.e8. doi: 10.1016/j.jpuro.2018.02.021.
- [39] Dicianno BE, Wilson R. Hospitalizations of Adults With Spina Bifida and Congenital Spinal Cord Anomalies. *Arch Phys Med Rehabil*. 2010; 91: 529-35. doi: 10.1016/j.apmr.2009.11.023.
- [40] Wilson R, Lewis SA, Dicianno BE. Targeted preventive care may be needed for adults with congenital spine anomalies. *PM R*. 2011 Aug; 3(8): 730-8. doi: 10.1016/j.pmrj.2011.05.021.
- [41] Filler G, Gharib M, Casier S, Lödige P, Ehrlich JHH, Dave S. Prevention of chronic kidney disease in spina bifida. *Int Urol Nephrol*. 2012; 44(3): 817-27. doi: 10.1007/s11255-010-9894-5.
- [42] Roth JD, Casey JT, Whittam BM, Szymanski KM, Kaefer M, Rink RC, et al. Complications and Outcomes of Pregnancy and Cesarean Delivery in Women With Neuropathic Bladder and Lower Urinary Tract Reconstruction. *Urology*. 2018; 114: 236-43. doi: 10.1016/j.urology.2017.11.052.
- [43] Szymanski KM, Cain MP, Whittam B, Kaefer M, Rink RC, Misseri R. All Incontinence is Not Created Equal: Impact of Urinary and Fecal Incontinence on Quality of Life in Adults with Spina Bifida. *J Urol*. 2017; 197(3): 885-91. doi: 10.1016/j.juro.2016.08.117.
- [44] Wiener JS, Suson KD, Castillo J, Routh JC, Tanaka S, Liu T, et al. Bowel management and continence in adults with spina bifida: Results from the National Spina Bifida Patient Registry 2009-15. *J Pediatr Rehabil Med*. 2017; 10: 335-43. doi: 10.3233/PRM-170466.
- [45] Oakeshott P, Hunt GM, Poulton A, Reid F. Expectation of life and unexpected death in open spina bifida: A 40-year complete, non-selective, longitudinal cohort study. *Dev Med Child Neurol*. 2010; 52: 749-53. doi: 10.1111/j.1469-8749.2009.03543.x.
- [46] Dicianno BE, Sherman A, Roehmer C, Zigler CK. Comorbidities Associated with Early Mortality in Adults with Spina Bifida. *Am J Phys Med Rehabil*. 2018; 97(12): 861-865. doi: 10.1097/PHM.0000000000000964.
- [47] Houtrow AJ, Maselli JH, Okumura MJ. Inpatient care for children, ages 1-20 years, with spina bifida in the United States. *J Pediatr Rehabil Med*. 2013; 6(2): 95-101. doi: 10.3233/PRM-130243.
- [48] Dicianno BE, Lovelace J, Peele P, Fassinger C, Houck P, Bursic A, et al. Effectiveness of a Wellness Program for Individuals With Spina Bifida and Spinal Cord Injury Within an Integrated Delivery System. *Arch Phys Med Rehabil*. 2016; 97(11): 1969-78. doi: 10.1016/j.apmr.2016.05.014.
- [49] Ekmark EM. Risky business: Preventing skin breakdown in children with spina bifida. *J Pediatr Rehabil Med*. 2009; 2(1): 37-50. doi: 10.3233/PRM-2009-0061.
- [50] Ouyang L, Bolen J, Valdez R, Joseph D, Baum MA, Thibadeau J. Characteristics and Survival of Patients with End Stage Renal Disease and Spina Bifida in the United States Renal Data System. *J Urol*. 2015; 193(2): 558-64.
- [51] US Preventive Services Task Force. The guide to clinical preventive services: recommendations of the US Preventive Services Task Force. Lippincott Williams & Wilkins; 2006.
- [52] Gaston H. Ophthalmic complications of spina bifida and hydrocephalus. *Eye (Lond)*. 1991; 5(3): 279-90. doi: 10.1038/eye.1991.44.
- [53] Froehlich-Grobe K, Nary DE, Van Sciver A, Lee J, Little TD. Measuring height without a stadiometer: Empirical investigation of four height estimates among wheelchair users. *Am J Phys Med Rehabil*. 2011; 90(8): 658-66. doi: 10.1097/PHM.0b013e31821f6eb2.
- [54] Dosa NP, Foley JT, Eckrich M, Woodall-Ruff D, Liptak GS. Obesity across the lifespan among persons with spina bifida. *Disabil Rehabil*. 2009; 31(11): 914-20. doi: 10.1080/09638280802356476.
- [55] Harris MB, Banta JV. Cost of skin care in the myelomeningocele population. *J Pediatr Orthop*. Jan 10 (3): 355-61. doi: 10.1097/01241398-199005000-00012.
- [56] Kim S, Ward E, Dicianno BE, Clayton GH, Sawin KJ, Beierwaltes P, et al. Factors Associated With Pressure Ulcers in

- Individuals With Spina Bifida. *Arch Phys Med Rehabil.* 2015; 1435-1441. e1. doi: 10.1016/j.apmr.2015.02.029.
- [57] Soe MM, Swanson ME, Bolen JC, Thibadeau JK, Johnson N. Health risk behaviors among young adults with spina bifida. *Dev Med Child Neurol.* 2012; 54(11): 1057-64. doi: 10.1111/j.1469-8749.2012.04402.x.
- [58] Hampton LE, Fletcher JM, Cirino P, Blaser S, Kramer LA, Dennis M. Neuropsychological profiles of children with aqueductal stenosis and Spina Bifida myelomeningocele. *J Int Neuropsychol Soc.* 2013; 19(2): 127-36. doi: 10.1017/S1355617712001117.
- [59] Cardenas DD, Topolski TD, White CJ, McLaughlin JF, Walker WO. Sexual functioning in adolescents and young adults with spina bifida. *Arch Phys Med Rehabil.* 2008 Jan; 89(1): 31-5. doi: 10.1016/j.apmr.2007.08.124.
- [60] Verhoef M, Barf HA, Vroeghe JA, Post MW, Van Asbeck FW, Gooskens RH, et al. Sex education, relationships, and sexuality in young adults with spina bifida. *Arch Phys Med Rehabil.* 2005; 86: 979-87. doi: 10.1016/j.apmr.2004.10.042.
- [61] Gamé X, Moscovici J, Guillotreau J, Roumiguié M, Rischmann P, Malavaud B. Sexual function of young women with myelomeningocele. *J Pediatr Urol.* 2014 Jun; 10(3): 418-23. doi: 10.1016/j.jpuro.2013.07.016.
- [62] Jackson AB, Mott PK. Reproductive health care for women with spina bifida. *ScientificWorldJournal.* 2007; 7: 1875-83. doi: 10.1100/tsw.2007.304.
- [63] Aldave G, Hansen D, Hwang SW, Moreno A, Briceño V, Jea A. Spinal column shortening for tethered cord syndrome associated with myelomeningocele, lumbosacral lipoma, and lipomyelomeningocele in children and young adults. *J Neurosurg Pediatr.* 2017; 19: 703-10. doi: 10.3171/2017.1.PEDS16533.
- [64] Messing-Jünger M, Röhrig A. Primary and secondary management of the Chiari II malformation in children with myelomeningocele. *Childs Nerv Syst.* 2013; 29(9): 1553-62. doi: 10.1007/s00381-013-2134-4.
- [65] Kirk VG, Morielli A, Brouillette RT. Sleep-disordered breathing in patients with myelomeningocele: the missed diagnosis. *Dev Med Child Neurol.* 1999; 41(1): 40-3. doi: 10.1017/s0012162299000079.
- [66] Snow-Lisy DC, Yerkes EB, Cheng EY. Update on Urological Management of Spina Bifida from Prenatal Diagnosis to Adulthood. *J Urol.* 2015; 194: 1-9. doi: 10.1016/j.juro.2015.03.107.
- [67] Eid AA, Badawy H, Elmissiry M, Foad A, Ebada M, Koraitim A. Prospective evaluation of the management of bowel dysfunction in children with neuropathic lower urinary tract dysfunction and its effect on bladder dynamics. *J Pediatr Surg.* 2019; 54(4): 805-8. doi: 10.1016/j.jpedsurg.2018.12.015.
- [68] Werhagen L, Hultling C, Borg K. Pain, especially neuropathic pain, in adults with spina bifida, and its relation to age, neurological level, completeness, gender and hydrocephalus. *J Rehabil Med.* 2010; 42(4): 374-6. doi: 10.2340/16501977-0529.
- [69] Dicianno BE, Kinback N, Bellin MH, Chaikind L, Buhari AM, Holmbeck GN, et al. Depressive symptoms in adults with spina bifida. *Rehabil Psychol.* 2015; 60(3): 246-53. doi: 10.1037/rep0000044.
- [70] McDonnell GV, McCann JP. Why do adults with spina bifida and hydrocephalus die? A clinic-based study. *Eur J Pediatr Surg.* 2000; 10(Suppl 1): 31-2. doi: 10.1055/s-2008-1072411.
- [71] Jernigan SC, Berry JG, Graham DA, Bauer SB, Karlin LI, Hobbs NM, et al. Risk factors of sudden death in young adult patients with myelomeningocele. *J Neurosurg Pediatr.* 2012; 9(2): 149-55. doi: 10.3171/2011.11.PEDS11269.
- [72] Marreiros H, Loff C, Calado E. Osteoporosis in paediatric patients with spina bifida. *J Spinal Cord Med.* 2012; 35(1): 9-21. doi: 10.1179/2045772311Y.0000000042.