Care for Adults with Spina Bifida: Current State and Future Directions

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The care for adults with spina bifida is an important area to study. As increasing numbers of patients with spina bifida survive into adulthood, they expect to thrive and receive the best possible care into adulthood to maintain their health. Understanding the health needs in this emerging and changing population will help clinicians provide the best anticipatory care for adults with spina bifida and continue to improve outcomes. This will also impact pediatric care by improving the ability to determine preventive methods from early on and understand the impacts of pediatric care and decisions over the lifespan. **Key words:** *adult, myelomeningocele, spina bifida, transition*

Spina bifida (SB) occurs in about 2.7 to 3.8 per 10,000 live births,[1] and more than 75% of people with SB live to adulthood.[2] Since the advent of antibiotics to treat surgery-related infections, great strides have been possible in surgical interventions with implantable devices. The ventriculo-peritoneal (VP) shunt to treat hydrocephalus was invented in the mid 1950s.[3] Since its introduction in the 1970s, clean intermittent catheterization (CIC) has improved the renal health and longevity for people with spinal cord lesions, reducing the risk of renal failure.[4-6] Optimizing neonatal surgery for infants born with SB has reduced early morbidity and mortality, and now people with SB live to adult years and participate in fulfilling lives. The new knowledge frontier is to optimize the health and community participation of people with chronic health conditions such as SB.

This article serves to provide an overview of the major health and psychosocial outcomes among the adult SB population and report on areas where data are needed. We begin by discussing the medical needs of adults with SB and outline the difficulties and benefits patients face when making the transition from pediatric to adult care. We then discuss the role of a national registry and the ways in which outcome measures could help improve the medical management of adult patients with SB. Finally, we outline the predominant areas of health that affect daily life of individuals with SB and propose that improved community participation should be one of the ultimate goals that guide clinical decision-making in patients with SB.

Medical Care Needs Into Adulthood

A need for coordinated care for adults with SB was described in 1994 by Kaufman et al, who demonstrated the increased occurrence of preventable conditions in a cohort of patients with SB after their multidisciplinary clinic disbanded.[7] This patient population was compared to a similar cohort in a clinic that continued to provide multidisciplinary care for adults. Data were collected from patient charts 3 years after closure of the clinic and revealed that 45% to 66% of the patients no longer received consistent care from primary care providers (PCPs) and specialists.[7] This was especially significant among the older patients. When compared to a multidisciplinary clinic that stayed intact, the patients with the least consistent care had more comorbidities than the control patients.[7] Even though the clinicians once seen by patients in this clinic were all still available after it disbanded, there was no coordinator of

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care. This resulted in a lack of follow-up care and missed treatments.[7] Issues of aging with SB may be compounded by the lack of coordinated care.

Lack of regular care by a PCP may result in increased preventable secondary complications and higher rates of emergency department (ED) admissions.[8-11] Previous research has reported increased rates of ED visits among adults with SB compared to the general population.[8]

Due to hospital or provider policy, insurance networks, or patient preference, people with SB may need to move to different providers or hospitals for their health care needs into adulthood. However, this transition may be met with resistance from patients and their parents as it can mean leaving multidisciplinary care clinics and instead receiving care from multiple adult specialists, who may not coordinate care or treatment decisions and may not have a specific knowledge or interest in treating SB.[8,12-15] One study found that the mean number of specialists seen by an adult with SB was 3.8 and that 71% of adults with SB reported difficulty in accessing medical care.[16]

Generally, transitioning to knowledgeable adult providers is appropriate and indicated for chronic childhood onset conditions, and there should be a planned, purposeful move to the new care provider or team.[17] Transitioning to care providers can also be difficult due to reduced family support, concerns with executive skills, and lack of access to knowledgeable adult care providers.[17]

Transition From Adolescent to Adulthood

Although some pediatric SB clinics continue to provide care for their patients into adulthood, many have age limits and patients must seek adult care elsewhere. Transition from adolescence to adulthood involves acquiring adult life skills and experience to manage health care tasks and decisions and developing and learning a new role as a recipient of care in an adult setting. A lack of motivation, reduced adherence in adolescence, or lack of clinical guidance on how to transition from long-term pediatric providers to adult care may cause patients to forego routine primary care or subspecialty appointments. Due to the range in ability levels and support systems among adults with SB, a one-size-fits-all model does not appropriately address all aspects of transitioning care.[14]

Resistance to the transition from pediatric to adult care is multifaceted and may come from anyone involved in an individual's care. Transition may be delayed because of anxiety about the change in providers or a lack of initiative due to apathetic feelings regarding such changes.[14] There may also be little incentive for patients to leave the safety and familiarity of the care and trusted relationships they have had their whole lives. Pediatric providers may feel resistance to terminate care of such patients, particularly due to lack of trust in and limited training of adult physicians in treating conditions that previously only affected children. Resistance may also come from parents who may feel that a transition to adult care would begin to exclude them from the dynamics of care and decision-making processes.[12]

On the other hand, it is possible that transitioning to adult care may provide patients with feelings of autonomy and independence in their abilities to manage care. Young adults who have made the transition describe feeling empowered to take responsibility in adult care settings.[12,14] Parents describe being surprised by their adult offspring's ability to step into the leading role, and they recognize the benefit in allowing this transition, despite their anxiety about their own loss of control.[18]

Transitioning to new providers can be facilitated by a coordinated approach that involves the pediatric care team and the family.[12,17] Discussions about the timing of the transfer of care should take place early among the patient, family, and pediatric provider.[12,17] This may help prepare the family and reduce their feelings of anxiety about making the transition in the future.[12] The Transition Readiness Assessment Questionnaire (TRAQ) may be a helpful tool for clinicians in determining the degree to which there is resistance to making the switch from pediatric to adult care.[15]

There is a documented difficulty with executive function skills amongst some individuals with SB when compared to the general population.[19] This may affect provider, parent, and patient readiness to transfer responsibility of important health care activities and decisions. One study found that among patients with SB, higher reported executive functioning was directly correlated with increased medical autonomy, when controlling for age, IQ score, and disability level.[19] Additionally, knowing the amount of support adults receive from their

support network may help clinicians better understand how to counsel young adults as they and their families proceed through the transfer of responsibilities for tasks and decision making to ensure the best outcomes.[12,19]

Role of Registries

The Centers for Disease Control and Prevention (CDC) registry for SB is a collaboration of 19 SB clinics across the country that collect data from all patients with SB to measure demographic information, treatments, and outcomes.[20] The registry is beginning to describe data elements at all ages and compare care and outcomes across centers. This information may help promote improvements in clinical pathways. For example, from previous cohort publications, much has been learned about morbidity and intellectual issues related to shunt malfunction and infection, resulting in efforts to reduce the frequency of shunt placement by 10% to 25%.[21,22]

There is a need to follow individuals with SB as they age in order to better elucidate the prevalence of specific complications. While current research cites correlations between functional ability and select outcomes, greater emphasis on potentially preventable secondary complications is warranted. Registries may be useful in filling in gaps in the knowledge of secondary complications among adults with SB and areas in which the clinical care they receive may be lacking. More literature is needed on both the prevalence of such secondary complications and evaluation of methods that attempt to treat or prevent and reduce these outcomes. Data collected from the registry may help clinicians guide their management of adult patients and may reveal areas where research is lacking. Such evaluations may also further improvements in pediatric care. For example, several studies have shown the benefits of a plantigrade positioned foot in reducing pressure ulcers, which can lead to osteomyelitis and partial amputation.[23-25] As these complications are areas of concern that occur among adults with SB, early intervention and orthopedic treatment of pediatric patients may improve these long-term health outcomes. Recognizing and understanding adverse health outcomes in adults with SB may thus inform treatment plans in pediatric care settings with the ultimate goal of improving long-term functioning.

Current Health Data Among Adults With SB

An extensive review of care for adults with SB by Dicianno et al highlighted areas where research is currently lacking.[26] As more individuals with SB are living later into adulthood, it is necessary to understand the frequency and type of care they receive in a given year to identify any gaps in their health management. Much research to date has focused on the association of lesion level with difficulties in multiple areas including cognition, ambulation, and bowel and bladder continence. Although this is useful as a baseline to compare or predict expected outcomes, these factors are not amenable to change at this time. Our recommendation is to shift the focus from such correlations and to instead identify outcomes that are amenable to change in order to improve health, functional quality of life, and community participation among patients. We will discuss current trends and cost of medical care among adults with SB and then highlight major areas that may be useful for clinicians in identifying secondary complications among this population.

Based on previous data regarding hospitalization rates among patients with SB, there is a need for increased awareness and monitoring of secondary complications. Individuals with SB have significantly more hospitalizations per year than the general population.[8,9,26] This finding was greatest among the patients between 18 and 39 years of age.[27] Data from a 2-year period (2004-2005) of adults 18 years of age and older with a secondary diagnosis of SB or associated conditions revealed that among 7,670 hospitalizations, 34% of the cases were due to potentially preventable secondary complications.[8] Of these cases of hospitalization due to preventable complications, 35.7% resulted in death.[8] Adults with SB were also reported to have greater than 2.5 times the mean number of 30-day long readmissions when compared to the general population.[27] One study reported 4 diagnoses associated with a higher likelihood of readmission for patients with SB regardless of age or gender: septicemia, chronic ulcer of skin, skin/subcutaneous tissue infection, and infective arthritis.[27]

One study demonstrated that patients with SB also have higher rates of inpatient admissions when compared to patients with cerebral palsy and traumatic brain injury.[10] This study sampled patients from a Canadian health care system where there is full public access to primary care, but there were no significant associations found between having a PCP and the number of inpatient care episodes. This may indicate a need for increased contact and communication with a knowledgeable PCP in order to prevent secondary complications.

Decreasing the rate of secondary complications could also significantly decrease medical costs. A previous study found the cost ratio of care for patients with SB compared to patients without SB to be 13.0 in children and adolescents ranging from 2 to 17 years of age. The cost ratio for adults aged 18 to 44 years old decreased to 6.8, however there are increased ongoing costs of medical care among this population.[28] PCPs can work with patients to identify areas of risk for complication, thereby preventing the need for hospitalization, decreasing costs, and improving overall health. For example, a study examining the prevalence and cost of inpatient admissions for urinary tract infections (UTIs) demonstrated 22.8 inpatient admissions per 1,000 patients with SB annually compared to 0.44 admissions per 1,000 patients without SB.[9] It is estimated that reducing the number of UTIs in patients with SB by 50% could decrease health care costs by approximately $4.4 million per 1,000 patients.[9]

There is a need for a comprehensive health care team for adults with SB, as these patients are at higher risk for health issues when compared to the general population. The primary aspects of general health concern in SB that we will be discussing include neurosurgical, urological, orthopedic/mobility, skin, metabolic, and psychosocial well-being (**Table 1**).

Table 1. Health factors affecting adults with spina bifida

Health factors	Monitoring methods	Outcomes
Neurosurgical care	Unknown, imaging Clinical serial muscle tests	Shunt malfunction Tethered cord syndrome
Urological care and renal health	Cystatin C level Creatinine level Renal and bladder ultrasound Culture for resistant organisms	Urinary tract infections (UTI) Kidney function Continence, amount of leaking Bladder, ureteral, renal stones
Cancer	Cystoscopy with biopsy Exams and cell testing (“pap”) Self exam, mammogram	Bladder cancer Cervical cancer Breast cancer
Hypertension	Mean arterial pressure, blood pressure	Vessel disease (atherosclerosis)
Metabolic syndrome	Fasting glucose level HbA1c level Lipid profile	
Obesity	Abdominal circumference (at umbilicus) Body mass index (BMI)	
Sleep apnea	Sleep study, symptoms questionnaires	Restful sleep, headaches
Skin breakdown	Pressure mapping	Decubitus ulcers Wounds
Pain	Pain questionnaires Pain medication use	Neuropathic Musculoskeletal Joint, overuse pain
Orthopedic care	X-rays	Fractures, hip dislocation Scoliosis
Bone health	Bone density scan (DEXA)	Fracture
Mental health	Questionnaire	Depression Anxiety
Sexual health	Questionnaire	Sexually transmitted diseases (STDs) Unplanned pregnancy Contraceptive use Sexual function

Shunt malfunction and neurosurgical care

Shunt malfunction is an area of key concern in adults with SB in relation to outcomes and mortality. Hydrocephalus is reported in 62% to 80% of adults with SB.[16,29,30] A national study of insured individuals with SB found complications from devices, grafts, or implants to be the second highest hospitalizing condition after UTIs.[8] Shunt revisions have been associated with poorer achievement in functional areas such as employment status, driving a car, and independent living.[31] Lack of achievement in these areas has been shown to be higher when shunt revisions occur beyond 2 years of age.[31]

The diagnosis of shunt malfunction can be difficult due to the overlap in symptoms between shunt malfunction and sleep apnea or depression. Understanding best diagnostic and treatment methods and outcomes is important to improve care of hydrocephalus. Symptoms of shunt malfunction may include headache, nausea, vomiting, and dizziness.[26,32] Drowsiness or lethargy appear to be common symptoms of shunt malfunction in long-term shunt-dependent patients. However, shunt symptoms in patients with chronic hydrocephalus can be subclinical and without significant or clear findings on imaging.[32] Unrecognized shunt malfunction may be a major underlying cause of death in adults with SB.[2] The registry may be helpful for determining the usual rates of shunt revision and the incidence of concomitant conditions such as sleep apnea or depression.

Diagnostic methods to identify sensitive symptom sets or findings are needed to identify which patients require surgical intervention. This is a key factor for families, who may have anxiety about neurosurgical transition. Radiological assessment should be performed to diagnose shunt malfunction; such imaging may still show small ventricles due to intermittent draining, ventricles with low compliance, and slit ventricle syndrome.[26] Therefore, it is helpful to have baseline imaging to serve as a comparison to future imaging. In some patients, imaging is not diagnostic and, if symptoms continue, revision may be clinically indicated to improve symptoms. It can be difficult to detect neurologic changes in someone with a disability at baseline, especially with a new provider.[33] The lack of specificity of these findings and the overlap with sleep apnea and depression highlight the importance of having a team or provider who knows the patient and his or her medical history.

As neurosurgeons may not routinely follow nonsymptomatic adult patients with SB at some centers, other key providers who know the patients and/or family well should monitor patients for shunt-related symptoms. A well-documented neurologic exam is key, as subtle signs may include brainstem findings of eye changes (optic nerve changes, changes in nystagmus or strabismus), swallowing issues, or new sleep apnea and snoring.

A Myelomeningocele Shunt Emergency Card has been developed and is free to print as a communication tool for a patient with a shunt (Ann and Robert H. Lurie Children's Hospital of Chicago website: https://www.luriechildrens.org/en-us/care-services/specialties-services/spina-bifida-center/resources/Documents/spina-bifida-emergency-card.pdf.)

Tethered cord syndrome

Tethered cord syndrome (TCS) results from tension on the spinal cord. It has been reported in 20% to 55% of patients with SB; increasing prevalence is positively correlated with the amount of time since repair.[26] A review study reported that adults with TCS typically present with back pain (57%-78%), motor weakness (44%-79%), and/or sphincter dysfunction (34%-71%).[34] The mean age of TCS onset reported among these patients ranged from 34 to 43 years of age.[34]

A longitudinal study also cited scoliosis, gait changes, and change in spasticity as the common preoperative symptoms in adult patients with TCS.[2] This study demonstrated that tethered cord release surgery improved or stabilized symptoms in 97% of a sample of 23 patients, whereas 3% of those patients reported worse symptoms post surgery.[2] More research is needed on the natural progression of TCS and the timing of intervention within the population of adults with SB. Frequency of this condition, risk factors, and optimal diagnosis and management could be better understood via a registry.

UTI

UTI may be the most common preventable secondary complication, and it has been reported in approximately half of adults with SB on an annual basis.[26,29] Methods to decrease this incidence rate should be tested, with reduction in antibiotic use as a secondary goal. Renal failure has been reported in 6% of one study population and each of these cases was preceded by multiple UTIs within a year, highlighting the importance of counseling patients on proper bladder management.[29] It is important to determine whether bacteriuria or cystitis (inflammation) is occurring, as some episodes may not need to be treated with antibiotics. Specific guidelines developed from registry information could help families and clinicians determine when antibiotics are necessary. Frequency of antibiotic-resistant bacterial growth is an important additional question that could be addressed via a registry.

A majority of adults (71%-85%) with SB are on CIC, and the majority of this subgroup is responsible for performing their own catheterizations.[2,35] The proportion of individuals who report continence is greater among adults on CIC versus adults not on CIC.[2]

A study on urological outcomes on adults with SB reported that 17% of a sample had surgery for stone disease. Of the surgeries, 57.4% were for renal stones, 31.1% for bladder stones, and 11.5% for ureteral stones.[35] Data on surgery for stone disease (bladder, ureteral, and renal) demonstrated that the majority of surgeries were performed on patients older than 18 years of age compared to pediatric patients younger than 18 years of age (80.4% vs 19.6%; $p < .001$).[35]

Monitoring of the long-term function of the bowel and bladder is warranted with anterograde cecostomy enema use, Mitrofanoff bladder stoma use, or other bladder and bowel regimens including ileocystoplasty, which shows high incidence of bladder calculi and renal scarring.[36] Registry comparisons between outcomes and complication rates using various bowel and bladder management methods may aid in understanding optimal management recommendations from childhood.

Cancer

Cancers may be underinvestigated in people with disabilities. One study comparing individuals with disability to the general population found that approximately 18% of individuals with disability had personal experience with cancer compared to only 7% of the general population.[37] Additionally, women with disabilities were 17% more likely to report noncompliance with gynecologic cervical exams and 13% more likely to report noncompliance with mammogram screening guidelines than women without disabilities.[37] Some pertinent barriers of adherence to cancer screening guidelines among individuals with disabilities include lack of health insurance and physical barriers at health care institutes in terms of mobility and getting into the correct position for screening procedures.[38,39]

Bladder cancer is rare in this population, however it can occur at an earlier age and present with different symptoms when compared to the general population.[40] Symptoms reported in one case series on 8 patients included gross hematuria, urosepsis, renal failure, difficulty catheterizing, increased frequency of UTI, and sterile pyuria.[40] Chronic inflammation poses a risk for the development of cancer, therefore patients with SB may be particularly at risk.[41] Median age of diagnosis of bladder cancer in this population is reported to be 37 years old, and 6 months is the median survival period.[40] Even though this diagnosis may be infrequent among individuals with SB, a high index of suspicion should be maintained with long-term urological care and immediate evaluation is necessary for any urological changes.[41] The rates of testicular cancer in this population are not described.

Hypertension

Reported rates of hypertension in adults with SB are between 5% and 25%.[16,29,35] There is a significant increase in the risk for prehypertension and hypertension with age in patients with SB compared to the general population [42] One study reported that fewer than half of the adults in a sample of patients with SB were considered normotensive. The reported rate of hypertension was 10.5% in adults with SB between the ages of 18 and 44 years old.[42] Rates of hypertension among this sample were directly correlated with age: 40.6% of adults 45 to 64 years old and 70.3% of adults 65 years or older.[42]

Rates of hypertension in adults with SB have been shown to correlate with obesity. Data from

one study of this population showed that 77% of obese adults had hypertension while only 26% of non-obese adults with SB were hypertensive.[16] Optimal treatment and outcomes of treatment of hypertension may be better understood via a registry.

Metabolic syndrome

Criteria for metabolic syndrome include waist circumference (>40 inches in men; >35 inches in women), blood pressure above 130/85 mm Hg, fasting triglyceride >150 mg/dL, fasting high-density lipoprotein cholesterol <40 mg/dL in men or <50 mg/dL in women, and fasting blood sugar over 100 mg/dL.[43] Rates of metabolic syndrome among individuals with SB (defined as having 3 or more risk factors) have been reported in a previous study to be 32% compared to 23% of the general population.[44,45] In this study, only 6% of patients with SB had zero risk factors of metabolic syndrome.[45]

Weight management and physical activity may be protective factors against developing metabolic syndrome. One study found that 76% of adolescents and young adults with SB were either inactive or extremely inactive.[46] Within this sample, physical activity was reported to be 50% lower than the general population and aerobic activity was 42% lower.[46] Adults with SB may have more barriers to routine physical activity such as medical complications, lack of social support, and limited accessibility to equipment and facilities suited for individuals with disabilities.[47] Due to the high prevalence of one or more risk factors for metabolic syndromes among patients with SB, it is necessary to provide counseling to these patients on lifestyle choices to prevent such risks and to determine most effective treatment/facilitation of lifestyle modulation and healthy lifestyle adoption.

Obesity rates among adults with SB have been reported between 30% and 37% in the literature.[16,46,48] Higher obesity rates have been reported among adults with public insurance compared to adults with private insurance.[48] Additionally, a higher proportion of adult women were considered extremely obese when compared to men.[48] Waist circumference and percentage of trunk fat may provide more accurate measurements of adiposity in this population than solely using body mass index (BMI) as a measure of obesity.[45,49]

The potential for diabetes and its complications and atherosclerotic disease associated with metabolic syndrome are concerning and should be monitored for via a registry.

Sleep apnea

Sleep apnea has been reported in approximately 10% to 20% of children with SB.[50-52] This may consist of obstructive, central, or mixed sleep apnea. In the general pediatric population, the rate of sleep apnea is about 4%.[50] Further research is needed to determine the prevalence of sleep apnea among adults with SB. Risk factors for patients with SB are different than risk factors in the general population. Chiari II malformation, lesion level, and use of a wheelchair for ambulation are some of the SB-specific factors that may increase risk for sleep disordered breathing.[50,51,53] Additionally, decreased strength in respiratory muscles or orthopedic conditions such as kyphosis and scoliosis may be associated with higher risk for restrictive lung disease.[51] Previous research has hypothesized that sleep apnea is often undiagnosed in patients with SB as it is not often assessed during clinical care.[51,53] Additionally, sleep apnea symptoms, such as morning headache, lethargy, and somnolence, overlap with depression and shunt malfunction, making the determination of optimal treatment difficult.[54]

Skin breakdown

Chronic skin ulcers are the primary diagnosis in approximately 6% of adults with SB, and skin and subcutaneous skin infections account for approximately 5% of hospitalizations among this group.[8] Acute hospitalizations for skin diseases and diseases of subcutaneous tissue were reported to be higher in adults compared to youth with SB (14.6% vs 7.4%).[11] One cohort study found that wounds were more prevalent in a teenage population than in adults 20 years old and older.[55] The foot and ankle were the most common location of wounds in adults, followed by the buttocks and the knee and thigh.[55]

Foot and ankle wounds are more prevalent in patients with a lumbar or sacral lesion level, whereas patients with thoracic level lesions

are more prone to wounds on the buttocks.[55] Pressure ulcers are more prevalent in males, wheelchair users, and patients with higher lesion levels, shunts, or bladder incontinence.[56] A registry could be helpful in identifying risk factors related to pressure ulcers on which to focus preventive efforts.

Pain

Pain has been reported in 25% to 32% of adults with SB.[57,58] In one study, approximately 10% of adult patients with SB had neuropathic pain, and the proportion of adults with neuropathic pain significantly increased with age (3% of adults 18-29 years of age, 13% of adults 30-40 years of age, and 29% over the age of 40 years).[57] Neuropathic pain was significantly higher among patients without hydrocephalus than with hydrocephalus.[57] Pain is a risk factor for depression and therefore should be assessed and monitored throughout adult care.[59] Neuropathic pain has been shown to have a significant impact on quality of life and happiness in other adult populations as well as in children with SB.[60,61] Registries could help clarify the effect of pain on mobility and health and identify optimal methods to reduce it.

Orthopedic care and mobility

Care issues in orthopedics include spine, back pain, scoliosis, walking, amputation, osteomyelitis, and osteoarthritis. Frequency of musculoskeletal complaints (shoulder, wrist, knee/hip, ankle/foot pain, and low back pain) and their impact on mobility are yet to be described for wheelchair users and individuals with SB using other assistive devices for mobility. Some related information exists in spinal cord injury populations.

A longitudinal study showed that patients who ambulated a majority (75%-100%) of the time during their teenage years continued to ambulate a majority of the time as young adults.[2] Lesion level may predict ambulation status into young adulthood, as previous studies have found a correlation between lower lesion level and greater proportion of time ambulating. [2] Understanding the factors that lead to loss of walking or conversely promote retention of mobility skills should be a goal for a registry.

Bone health

It has been reported that bone mineral density is reduced in patients with SB compared to typically developing peers.[62,63] One study found that children and young adults with SB have decreased levels of vitamin D, and hypophosphatemia as a result, when compared to the general population.[63] Risk factors among adults with SB include renal failure, use of epilepsy medications, and decreased physical activity.[26] Similar to the general population, female adults with SB are at an increased risk compared to males.[26] Decreased bone mineral density is predictive of an increased risk for fracture within this population.[63] One study suggests increased supplementation, particularly for vitamin D, is necessary in this population. However, guidelines have not been published in terms of effective dose and the age at which to begin supplementation.[26,63]

Mental health

Individuals with SB have higher rates of anxiety and depression compared to the general population. It is important to assess the mental health status of patients making a transition from a multidisciplinary pediatric model of care into adult care. Rates of depression among adults with SB have been reported to be between 25% and 33%, while rates of anxiety have been reported as 13%.[59,64] Thirty-six percent of adults with SB in one study were on antidepressants to treat depression and 63% of the individuals with depressive symptoms were on antidepressants.[64] No gender differences are reported in terms of anxiety and depression.[59] A positive attitude toward SB and satisfaction with family functioning have been reported as modifying factors to depression, while pain has been reported as a risk factor for both depression and anxiety.[59] Depressive symptoms may go unrecognized among adults with SB, and they should be screened for these symptoms during clinic visits to ensure proper treatment. More research is needed to determine frequency of mental health symptoms and effective

interventions to treat depression among adults with SB.

Sexuality and health education

In addition to medical complications, there are other health outcomes that should be addressed in the clinical setting such as sexual function and healthy pregnancy. One study found that although adults with SB obtain information on sexuality from a variety of sources, information on sexual function as it relates to SB primarily comes from health care providers.[65] Among this study sample, 82% knew that sexually active patients with SB need to use a form of birth control and 96% knew sexual activity poses a risk for sexually transmitted diseases (STDs).[65] However, only 70% knew that females with SB could get pregnant, 72% knew that condoms should be latex free, and 62% knew of the need for folic acid supplementation prior to conception.[65] Additionally, data have shown that 24% to 35% of patients with SB reported that they have had sexual intercourse.[65-67] Therefore, it is necessary to provide sexual health education during adolescence and adulthood to improve patient knowledge and adherence to health-conscious practices.

Erectile function and fertility for men with SB also need to be understood to ensure counseling and treatment for healthy sexual function. One study found that erectile dysfunction could not be predicted solely by the level of neurologic lesion.[68] Twenty-three percent of patients in this study were found to have erectile dysfunction, but none of them were being treated for it.[68] More research is necessary to further evaluate sexual function in adult males with SB.

Birth control counseling should be offered. Women with SB may have higher risk for adverse effects of oral contraceptives and intrauterine devices (IUDs). Oral contraceptives that are mostly progesterone may put women at a higher risk for osteoporosis, while contraceptives that are estrogen based may increase risk of hypercoagulability, particularly in persons with decreased mobility.[69] Additionally, IUDs should not be used in patients with chronic UTIs, diminished pelvic sensation, or prolapsed uterus as these conditions may increase the risk of infection.[69] Information about outcomes and complications of use of various forms of contraception is needed.

Risks of adults with SB having a child with SB are increased compared to the general population (4% vs 0.1-0.3%).[70] Females of childbearing age in the general population require folic acid supplementation to reduce the risk of neural tube defects in pregnancy[71]; adults with SB may require higher doses of folic acid to achieve the same preventative outcomes.[69] The CDC recommends a dosage of 4 mg/day of folic acid 1 month prior to trying to get pregnant, while a different study recommended a dosage of 5 mg/day 2 months prior to conception for women with neural tube defects or with a family history of neural tube defects.[72,73] Women at increased risk for having a child with neural tube defects should adhere to these recommendations of increased folic acid compared to the general population and should continue these dosing recommendations through the first trimester.[72,73] Counseling should include advice about folic acid supplementation for the patient or their partner. Additionally, female first-degree relatives or partners of male first-degree relatives should be made aware of the importance of high-dose folic acid supplementation prior to conception or during fertile years.[74]

Community Participation

A study by Bellin et al[30] used variables under the "activities and participation" domain of the International Classification of Functioning, Disability and Health framework to assess community living in adults with myelomeningocele. Reports on self-management were similar between genders and were predictive of independent living but not of employment.[30] It was found that males are more likely to gain employment and females are more likely to live independently.[30] Reports of employment rates among adults with SB have been cited between 23% and 42%, and rates of independent living have been cited between 13% and 24%.[30,59] Rates of completion of high school or high school equivalence (GED) among individuals with SB have been reported in 94% of one study population, and completion of a technical,

associates, or college degree was reported in 62% of this sample.[75] Another longitudinal study following adults with SB found that 85% were enrolled in high school or college or recently graduated.[2] Education in a typical classroom setting has been reported in 62% to 63% of students; other students may require special education classes or individual assistance.[2,75] It is important for clinicians to keep domains such as employment, community participation, and independent living in mind in order to make recommendations to patients on counseling services and training programs that may help to promote their autonomy and improve quality of life.

Peer relationships are another domain that may be predictive of quality of life and community participation in adults with SB. One study found that emerging adults (18 and 19 years of age) with SB reported a similar number of friends as their typically developing peers.[76] Lower levels of executive dysfunction were found to be predictive of a greater number of friends.[76] Research on peer relationships in individuals with SB has primarily focused on youth and adolescent stages. More research is needed to investigate the number of friends in adults with SB and the quality of those relationships.

Conclusion

There are many factors that affect the health and well-being of people with chronic conditions such as SB. The goal of treatment at this junction is to improve health outcomes in SB, as well as to improve quality of life and community participation as the ultimate goal. We know that community participation has a greater association with quality of life and life satisfaction than individual health outcome aspects. Improving health and health care costs for people with SB would allow for maximum integration into work, community living, and social-recreational activities in the community. Although maintaining appropriate health care monitoring and providing timely preventive and episodic care is important, it should be done in the context of improving quality of life and functioning. It is helpful to structure patient assessments and recommendations within the World Health Organization's International Classification of Functioning Disability and Health framework. In using this structure, medical care can be focused toward creating protocols to treat comorbidities that have the potential for improvement and impact participation.

Registries can have a key role in understanding the natural history of a chronic condition such as SB and assess ways to reduce complications and provide better anticipatory care. Understanding outcomes in adulthood can affect care delivery in pediatrics; early care can be provided to reduce unwanted outcomes. Future research should focus on such modifiable outcomes that improve the health and quality of life for these patients, through all stages of life. Registries and identified patient-centered outcomes will be key in identifying research goals to achieve these goals in the years ahead.

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