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Spina Bifida Guideline

Neuropsychological care guidelines for people with spina bifida

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Abstract. While the neuropsychological profile for individuals with Spina Bifida (SB) can vary, often certain patterns of strengths and weaknesses are evident across the lifespan. Understanding variability related to neural structure, genetics, ethnicity, and the environment is key to understanding individual differences in outcomes and can be vital in planning interventions and tracking progress. This article outlines the SB Guideline for the Neuropsychological Care of People with Spina Bifida from the 2018 Spina Bifida Association's Fourth Edition of the Guidelines for the Care of People with Spina Bifida and acknowledges that further research in SB neurocognitive profiles is warranted.

Keywords: Spina bifida, myelomeningocele, neuropsychology, guidelines, developmental disabilities, neural tube defects

1. Introduction

Neuropsychological studies show a pattern of strengths and weaknesses involving motor, cognitive, academic, and adaptive functions in individuals with Spina Bifida [1–3]. This pattern is most commonly seen among individuals with Spina Bifida who are born with an open spinal lesion (myelomeningocele) and usually have a Chiari II malformation and other congenital brain malformations involving the cerebellum midbrain, and corpus callosum [4]. Most of the existing literature is based on patients who had hydrocephalus treated with surgical implantation of a shunt; however, the literature is just emerging on younger populations with myelomeningocele treated for hydrocephalus with different interventions, such as endoscopic third ventriculostomies Prenatal surgery is also preventing hydrocephalus and the need for shunting in some patients. It remains to be seen whether these different treatment modalities are associated with different neurocognitive profiles.

It is important to identify this subgroup of patients with myelomeningocele, which makes up 9% of the population with Spina Bifida. Individuals born with other types of Spina Bifida do not have the changes in neuroanatomical development referred to above and often have more typical cognitive development [5]. The Spina Bifida Myelomeningocele (SBM) neurocognitive pattern involves strengths in learning skills and performing tasks that rely on associative, rule-based processing (e.g., math fact retrieval, word reading), and weaknesses when learning and performance involves the construction or integration of information (e.g., math problem-solving, reading comprehension) [2].

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To illustrate, children with SBM have difficulty with controlled motor performance tasks that require adaptive matching of a motor response to changing visual information which involves the cerebellum [2], and is associated with the Chiari II malformation. However they can learn motor skills through repetition and correction of errors [6–10] which involves the relatively preserved basal ganglia [4].

In language and reading areas, vocabulary, grammar, and word recognition are strengths [11]. However, children with SBM experience challenges in listening and reading comprehension, and in oral discourse involving the use of language in context (pragmatics) [12–14]. This has been linked to anatomic changes in the corpus callosum [15].

In mathematics, children with SBM can learn math facts; however, complex procedures that require multiple steps and algorithms are an area of challenge. They often experience difficulties with estimating quantities and have impaired math problem-solving skills [16–18]. Problems with math a longterm predictor of adult independence, are common in adults and children with SBM [2,19].

Many children with SBM meet criteria for Attention Deficit/Hyperactivity Disorder, Predominantly Inattentive Presentation (ADHD) [20,21]. However, in contrast to children with developmental forms of ADHD related to self-regulation, the attention profile of children with SBM is characterized by under-arousal and excessive persistence in controlling attentional focus. These difficulties in alerting and orienting to external stimuli are related to disruptions in midbrain and posterior cortex and are discernable from infancy [22,24]. It is more appropriate to conceptualize attention deficits, for patients with SBM, as involving posterior brain pathways; this is in contrast to the "executive dysfunction" model, such as with developmental ADHD, traumatic brain injury, and other disorders involving frontal lobe functions.

Despite the modal neuropsychological SBM profile presented above, there is a great deal of variability in neuropsychological outcomes. Understanding the variability in neuroanatomic anomalies, ethnicity, and the environment (socio-economic status and education) is key to understanding individual (rather than group) differences in outcomes. Neurological status, including more severe hydrocephalus, repeated shunt malfunctions, and ethnicity predict poorer outcomes and deviation from the modal profile [5]. Individuals with higher lesion levels have more severe neuroanatomic brain malformations and higher rates of intellectual disability. Spinal defects at T12 and above are more frequent among individuals of Latinx ethnicity, likely related in part, to genes involved in folate metabolism [5,25]. These populations also are often economically disadvantaged, with diminished access to care and adverse outcomes attributable to social determinants of health, further increasing risk for negative neuropsychological outcomes [5]. These disadvantaged children may not display the aforementioned relative strengths in language and academics that are more typical for children with SBM with lower level lesions and equitable socio-economic circumstances.

2. Guidelines, goals and outcomes

The goals of the neuropsychology guidelines were both practical and aspirational. Although neuropsychological assessment is clearly indicated for most patients born with SBM, barriers to care include limited access to clinicians with experience in the evaluation of complex medical patients and access to resources (and/or adequate insurance coverage). This process was initiated through the creation of several goals and desired outcomes:

Primary

1. Optimal development of language academic, and other learning skills.
2. Optimal performance in school, university, and vocational settings.

Secondary

1. Maximize independence according to individual capabilities.
2. Maximize participation in society.

Tertiary

1. Acquisition of a job.
2. Utilization of learning skills is apparent in a variety of contexts.

3. Methods

The process of the guideline development was set by the Executive Committee created by the Spina Bifida Association of America (SBA). It included a mix of providers who have worked with patients who have Spina Bifida throughout their careers as well as SBA professionals. The process began with a level 1 review of the literature, updated to include studies that were published between 2002 through 2015, which resulted in the review of 2449 abstracts; these were then for-

Table 1
Clinical questions that informed the neuropsychology guidelines

Age group (from guidelines)	Clinical questions
0–11 months	1. What early interventions in infancy are appropriate for supporting the development of motor, cognitive, and early literacy and numeracy skills? 2. How are new treatments such as prenatal repair in the Management of Myelomeningocele Study (MOMS) and the Endoscopic Third Ventriculostomy/Choroid Plexus Cauterization (ETV/CPC) affecting the health and development of infants? 3. How can teams use early Magnetic Resonance Imaging (MRI) findings (eg, malformations dysplasia, reduced volume, and agenesis) to predict domains of risk and identify potential early interventions to support development?
1–2 years 11 months	1. What early interventions are appropriate for supporting the development of motor, cognitive, and early literacy and numeracy skills? 2. How is the health and development of children changing with prenatal surgery (MOMS trials)? 3. How is the health and development of children changing with the use of new surgical procedures such as ETV/CPC rather than shunting? 4. How does monitoring for hydrocephalus and delayed shunting alter development?
3–5 years 11 months	1. How does the relationship between the nervous system and mental functions among children with SBM affect their learning in reading, mathematics writing social science, and science? How does it affect them at different developmental stages? 2. What do teachers, psychologists and other professionals need to know about the development of individuals with SBM?
6–12 years 11 months	1. How does the relationship between the nervous system and mental functions among children with SBM affect their learning in reading mathematics writing social science, and science? How does it affect them at different developmental stages? 2. What interventions support their cognitive development and academic achievement? 3. What do teachers, psychologists and other professionals need to know about the development of individuals with SBM?
13–17 years 11 months	1. How does the relationship between the nervous system and mental functions among individuals with SBM affect their learning in reading, mathematics writing social science, and science? How does it affect them at different developmental stages? 2. What interventions and programs provide smooth transitions to post-secondary education and/or career and vocational training? 3. What do teachers, psychologists and other professionals need to know about the development of people with SBM? 4. How do treatment teams help prepare their patients for the transition to adulthood and to take on their own medical care? What indicators are helpful to a team in identifying individuals who may require ongoing support in order to have adequate management of their medical conditions?
18+ years	1. How do treatment teams help prepare all of their patients for the transition to adulthood and the assumption of their own medical care? 2. What indicators are helpful to a team in identifying those who may require ongoing support for adequate management of their medical conditions?

warded to the level 2 review that resulted in 874 full text articles being archived for working groups. Working groups then reviewed the literature that was forwarded to each committee, and were able to add and subtract research articles based upon their contribution to the field. The full guidelines then included 803 articles; the neuropsychology working group cited 4 articles in their guidelines. The teams worked to craft goals and potential outcomes, along with specific clinical questions for each of the six age groups (Table 1), and then to systematically evaluate the literature's documentation of the 'answers' to the clinical questions. Following peer review by two sets of colleagues, including those in fields working in close collaboration with neuropsychology (e.g., Neurosurgery, Psychology, Transition, and Family Functioning groups) and a broader set of multidisciplinary colleagues, the final version of the guidelines was published in 2018 [26]. The full methodology followed by the working group for writing these guidelines was described by Dicianno et al. [27].

4. Results

Using the existing literature, as well as our own clinical consensus, the guidelines were developed (Table 2). They document that neuropsychological assessment is indicated throughout the lifespan. Key developmental guidelines included early elementary school in order to identify changes in learning; and the transition out

Table 2
Neuropsychology guidelines

Age group	Guidelines	Evidence
0–11 months	1. Provide parents with formal teaching and intervention around the development of effective parenting practices for fostering developmentally appropriate and responsive parent-child interactions.	[28], clinical consensus
	2. Teach parents more interactive parenting strategies, as research has shown that doing so in infancy results in significantly stronger cognitive and social language outcomes (at age 3) and better social problem-solving skills (at age 7).	[6, 29]
	3. Closely monitor infants who have undergone prenatal treatment, given the paucity of literature on their long-term outcomes.	[30, 45]
	4. Use infant development scales that assess cognition, language, motor, and social development for all infants with SBM, including those who have not been surgically treated for hydrocephalus.	[31]
	5. Adaptive behavior assessments that are interview-based are easy to complete and sensitive to growth trajectories in development.	[3]
1–2 years 11 months	1. Monitor and evaluate onset and progression of physical, cognitive, communicative, and social development; refer all children in this age group with SBM to an early intervention program. If children are discharged or are receiving private services, any changes in development warrant a re-referral to a formal program for early intervention/birth-three years.	Clinical consensus, [28, 31]
	2. Implement parental involvement with formal early intervention supports for both: – language (e.g. delayed onset, articulation difficulties, or unusual patterns of development such as excessive imitation, difficulties in language comprehension); – physical and occupational therapy for independent mobility, strengthening	Clinical consensus
	3. Teach and encourage parents to engage in effective interactions that facilitate the child's movement and exploration, language and communication, and play. Children of parents with higher expectations who facilitate attention, require movement, and support language development have better outcomes.	[6, 28–29], Family Functioning Guidelines [26]
	4. Encourage the use of equipment that facilitates object exploration and manipulation because it can be essential in providing access to their environment. This may include seating to support the trunk with a large enough tray to catch objects that are dropped and parent assistance with maintaining attention to objects that are able to be manipulated and explored by the child. These supports can often be obtained through early intervention programs/birth to three, occupational/physical therapy services, or physiatry.	Clinical consensus, Mobility Guidelines [26]
	5. Provide encouragement to participate in group learning experiences, especially when families are unable to find available day care that attends to necessary medical needs. These group learning experiences can be provided through either community groups or early intervention.	Clinical consensus
	6. Monitor developmental progress based on thorough assessments beyond determination of milestones, which are weak indicators of developmental difficulties. Shifts in the rate of skill development and skill regressions can reflect changes in medical status that warrant urgent follow up.	Clinical consensus
	7. Conduct periodic assessments with age-appropriate measures of early language skills because these can help identify more subtle difficulties of development.	[31]
	8. Monitor coordinated upper limb movement and attention multiple times per year in children with severe Chiari malformation, tectal beaking, and callosal hypogenesis.	[9, 24]
3–5 years 11 months	1. It is essential to carefully monitor the development of attention and self-regulation skills; these begin to emerge as a separate domain and directly affect the subsequent development of cognitive, academic, and social skills. Expectations for independent problem solving, responsibility, and social interactions are critical for school performance and psychosocial adjustment. Preschoolers with SBM show early manifestations of attention, pragmatic language, and math difficulties that subsequently emerge as major factors in academic and social adjustment.	[2, 12, 29, 32]
	2. Patients with identified concerns, even if mild, require timely referrals to the local special education preschool program and/or outpatient providers (e.g., psychologist, developmental pediatrician).	Clinical consensus
	3. Monitor language comprehension problems because interventions may facilitate the development of vocabulary and conversational speech that are essential for reading comprehension later in school.	[3, 12, 31]

Table 2, continued

Age group	Guidelines	Evidence
	4. Carefully observe children with more severe hydrocephalus, hypogenesis and/or severe hypoplasia of the corpus callosum and history of central nervous system infection because they are at greater risk for difficulties involving construction of meaning from language. These skills need to be carefully tracked by preschool education teams or through formal assessments with neuropsychologists, developmental specialists, or speech and language pathologists.	Clinical consensus, [15, 31, 33]
	5. As part of the child's medical team, advocate for access to high quality public education with related services that support the development of attention, self-regulation, social interaction skills, and independence. If parents choose private school or decide to home school, then formal assessments and recommendations for support services and supplemental resources should be provided in those settings as well. All children, regardless of placement, can and should be evaluated for eligibility for special educational services when learning problems are present.	clinical consensus
	6. Use "other health impaired" classification (or neurological disorder classifications in some states) help schools understand that potential learning difficulties are related to the underlying neurological disorder. Help all individuals who interact with the child understand that SBM is not simply an orthopedic condition or "physical disability." Brain malformations and hydrocephalus (with or without shunting) affect learning, especially in areas that require the construction and integration of information such as language, reading comprehension, and mathematics.	[3, 5, 11–14, 16–19, 32, 39]
	7. Monitor development with early math and literacy skills assessments to help establish an understanding of more subtle developmental difficulties and the need for more tailored educational supports.	[11–14, 16–19, 31–32, 39]
	8. When available, consider full neuropsychological evaluations that include the assessment of early literacy and numeracy skills. Neuropsychological assessments provide a more comprehensive understanding of strength and weakness, as well as significant discrepancies that may not be captured by psycho-educational testing that is provided by school districts.	Clinical consensus
6–12 years 11 months	1. Orient health care professionals that an individual with SBM does not simply have an orthopedic impairment. Explain to them that brain malformations and hydrocephalus (with or without shunting) affect learning, especially in areas that require the construction and integration of information such as self-management skills. Learning is facilitated when it is based on rules that can be verbally mediated and rehearsed, much like a recipe. This is especially important for bladder and bowel interventions for which the child's participation at an early age facilitates independence and social adjustment along with adherence to dietary regimens.	[22, 41, 44]
	2. Abstract concepts and global guidelines about self-care are ineffective for skill acquisition. It is essential to create routines, so that practice and repetition of self-management tasks can become rote activities. Coach clinical teams to carefully formulate instructions to be verbally mediated and to emphasize rule-based learning with repetition and rehearsal.	Clinical consensus, Health Promotion and Preventive Services Guidelines, Nutrition and Obesity Guidelines [26]
	3. Orient educators and school-based professionals that an individual with SBM does not simply have an orthopedic impairment and that brain malformations and hydrocephalus (with or without shunting) affect learning, especially in areas that require the construction and integration of information such as language, reading comprehension, and math problem-solving. Psycho-educational assessments can track global intellectual and academic progression, but they rarely assess the development of essential skills in attention, executive functioning, coordinated upper limb, and memory domains, as well as adaptive skill acquisition. Children with SBM benefit from a full neuropsychological assessment, when available.	Clinical consensus, [2, 11–14, 16–19, 21, 32, 38–40, 44, 46]
	4. Monitor school age children carefully for the onset of academic, attention, and behavioral difficulties. These problems tend to be identified later in school, partly because of the early development of word recognition, rote numerical skills, and vocabulary skills (usually in children who are not from socially- and economically- disadvantaged settings) that mask the presence of difficulties with math and reading comprehension.	Clinical consensus, [2–3, 5, 11–14, 16–19, 21. 32. 38–40, 44, 46]
	5. Carefully monitor children for the onset of attention problems, as they are often interpreted as motivational or behavioral issues and are manifested as lack of focus, slow cognitive tempo, failure to initiate, and infrequently with hyperactivity or impulsivity.	[21, 38, 46]
	6. Attention problems are correlated with the Chiari malformation, tectal beaking, and hypogenesis of the corpus callosum.	[22–24, 33, 35–36]

Table 2, continued

Age group	Guidelines	Evidence
	7. Follow American Academy of Pediatrics guidelines when evaluating for ADHD. One-third of individuals meet the criteria for ADHD, Predominantly Inattentive Presentation on parent rating scales.	[5, 31, 38, 43]
	8. Interventions for attention problems that involve medications may be tried, but clinical experience suggests that lower doses are effective and that many with SBM do not respond robustly to stimulants, most likely because the underlying attention problem emerges from posterior components of the attention network and not from the frontal-striatal networks (as in developmental ADHD).	Clinical consensus, [3, 23, 38, 46]
	9. Monitor for the development of language and reading problems. The severity of hydrocephalus and corpus callosum malformations affects the child's ability to integrate information and to construct meaning from language.	[2, 11, 12–13, 15, 33]
	10. Over 25% of children with SBM have significant language and reading comprehension problems which tend to be present both for listening and reading comprehension.	[3, 11, 13–14]
	11. Because of these common academic difficulties in children with SBM, formal assessment should include text-level reading comprehension and not just word reading accuracy and fluency.	[11, 13–14]
	12. Monitor children for the development of math problems. Over 50% of children with SBM develop math difficulties.	[5, 16–19, 32, 39]
	13. Assessment of mathematics should include assessment of complex calculation skills and, in the later grade school years, math word problems.	[16–19, 32, 39]
	14. Implement interventions like those used with children with learning disabilities when a child has a problem with reading or math, as these are often effective. For example, although problems with word reading and phonological awareness are rare in those with SBM, treatment programs like those used with children with dyslexia have been shown to be effective. Another example is the successful use of math problem-solving interventions designed for those with math disabilities. Take advantage of their strength in rule-based learning by providing explicit, well-structured instruction.	[3, 40]
	15. Use assistive devices as early as possible when developing writing programs. Keyboarding is a viable alternative to handwriting, although some practice with paper and pencil skills is useful through most of elementary school. Keyboarding must be taught and rehearsed if it is to be useful. Accommodations for writing difficulties are critical components of the educational plan.	Clinical consensus
	16. When available, consider full neuropsychological evaluations that include the assessment of early literacy and numeracy skills. Neuropsychological assessments provide a more comprehensive understanding of strength and weakness, as well as significant discrepancies that may not be captured by psycho-educational testing that is provided by school districts.	Clinical consensus, 3
13–17 years 11 months	1. Promote interventions that address integration and assimilation of information with a specific focus on reading comprehension and mathematics problem-solving, as well as other areas of applied mathematics and functional numeracy.	[5, 11, 16–19, 32, 39]
	2. Intervention programs should be maintained because their absence is associated with plateaus in skill development in most populations with disabilities.	[40]
	3. Encourage participation in school-related and extracurricular activities Create vocational plans and transitional services with a focus on functional independence.	Clinical consensus
	4. For students receiving special education services, the Individualized Education Plan (IEP) is required to include a formal transition plan to address vocational, occupational, and life skill domains by 14 to 16 years of age. Coach parents to ask about educational transition plans and to request evaluations to bolster the plans. Early transition plans are essential to develop the capacity to assume the roles and responsibilities of the post high school environment and achieve optimal independence. They are also needed to ensure that appropriate referrals are made to adult agencies, that there is suitable life and vocational skill training, and that there are discussions about plans after high school. Educate families about the need for a transition plan and check to ensure a comprehensive plan is created. If needed, refer to state-based educational advocacy programs (e.g., the ARC) that can provide support and education.	Transition Guidelines [26]
	5. Because social skills of individuals with SBM are strongly related to neuropsychological variables, namely attention and executive function, consider using psycho-educational and/or neuropsychological assessments to inform psychosocial interventions and mental health supports.	[29, 34, 42], Mental Health Guidelines [26]

Table 2, continued

Age group	Guidelines	Evidence
	6. Be aware that in addition to the cognitive and learning problems associated with the underlying neurological disorder, persons with SBM may experience reduced quantity and quality of social interactions. Encourage structured opportunities for social interaction through school, church, and afterschool opportunities.	Clinical consensus
	7. Conduct yearly screening and timely referrals for appropriate diagnosis and treatment of anxiety and/or depression with psychotherapy and/or medication treatment as needed.	[42], Mental Health Guidelines, Quality of Life Guidelines [26]
	8. Identify cognitive strengths and weaknesses for those who are assuming responsibility for their own medical care. This may require formal assessment, particularly if children are unable to assume responsibility for their own medical decisionmaking and will require guardianship. Efforts to assess and build communication skills, increase knowledge about their medical condition and history, and develop medical triaging skills needs to begin as early as possible because it may take those in this age group over several years to learn the skills necessary to understand and take responsibility for their own medical care. Address bladder and bowel incontinence, as both can be major issues affecting social adjustment.	Clinical consensus [37 41, 45], Bowel Function and Care Guidelines, Urology Guidelines, Transition Guidelines [26]
	9. Advise children and/or their parents/guardians to obtain copies of psycho-educational and/or neuropsychological assessments. Explain that documentation of an intellectual disability and/or learning disability prior to age 18 is needed to qualify for services in adulthood. A diagnosis of intellectual disability requires thorough assessment of adaptive skills. This is an important point because school programs and special education service evaluations may not always include formal assessment of adaptive skills.	[40, 43]
18+ years	1. Many patients with intellectual disabilities or significant learning challenges will remain eligible for services through their local school districts until 21 or 22 years of age. When young adults are eligible, these services provide access to both vocational and lifeskills training that are essential to support the development of stronger functional independence skills.	Clinical consensus, [43]
	2. Encourage that vocational services addressing job skills, additional education, and related activities be provided to appropriate individuals in a timely manner. Referrals to state-based agencies are commonly included in transition programs, and found in special education documentation/IEPs.	Clinical consensus
	3. For students who received special education (IEP) or 504 Plan accommodations in high school, ongoing supports under the Americans with Disabilities Act (ADA)/Section 504 of the Rehabilitation Act are necessary. For those attending college, refer them to their college's office of disability services for ongoing educational supports. Many students will also require an updated neuropsychological assessment to support eligibility. For those in workplace environments, refer to the state-based rehabilitation/vocational commission for additional support.	Clinical consensus
	4. In preparation for the transition to adult care models, where often times less coordination of medical care is provided, medical team members must take an active teaching and training role to build the necessary skills to support transition. Teach the patient the skills necessary to effectively communicate with staff, recognizing that they may prefer a different method than their parents (e.g., phone calls vs. internet portal). Test on important aspects of their medical conditions, regimens, and allergies. Rehearse triaging medical symptomology, with clear guidelines on when to seek medical care, to mastery (e.g., not when they first get it right, but when they always get it right).	Clinical consensus, Self-Management and Independence Guidelines, Transition Guidelines [26]
	5. Continuously monitor cognitive skills, especially math, memory, and attention, to ensure the maintenance of learning skills essential for work and independence.	[19, 21,22 44]
	6. Changes in these areas may be a sign of unidentified shunt failure or shunt dependency, or other significant medical problem requiring intervention.	Clinical consensus
	7. Full neuropsychological assessment is recommended for adults with SBM who experience cognitive decline and suspected shunt failure.	Clinical consensus
	8. Monitor for mental health concerns and potential cognitive decline with aging.	Mental Health Guidelines [26]

of high school to assess for independent functioning levels.

Although there is often a typical profile of strengths and weaknesses, there is variability in the profile and level of severity of neuropsychological functioning. In addition, multiple factors, including health status and medical history, as well as family structure and access to intervention, impact both the acquisition of skills over time and overall adaptation. The guidelines therefore recommend neuropsychological consultation, monitoring, and evaluations throughout the lifespan.

Following the review of the literature and the creation

of the guidelines, there were still some unanswered clinical questions that support the need for ongoing research and updating of guidelines in the future. Additional research was considered essential given the changes in neurosurgical care that are currently occurring with prenatal surgery and endoscopic third ventriculostomy and choroid plexus cauterization procedures (ETV/CPC) procedures. However, the need for additional research on the aging brain was also highlighted.

5. Discussion

The SBA Guidelines for the Neuropsychological Care of People with Spina Bifida recommend neuropsychological evaluation at key times to identify strategies for learning and improve outcomes for independent functioning. Evaluation is also recommended in order to better delineate individual variations in the SBM modal profile. Lastly neuropsychological assessment is recommended in order to better understand impact of new neurosurgical interventions.

5.1. Early childhood

Understanding the basis for strengths and weaknesses in SBM and using that information to inform interventions may prevent some of the negative outcomes. Fletcher et al. [3] suggest four areas that may be important for facilitating the early development of children with SBM, including (a) early movement, with a focus on encouraging the child to initiate and respond to environmental contingencies that require action [7]; (b) early language, to ensure development not only of vocabulary and literal language skills, but the child's sensitivity to contextual and pragmatic aspects of language [12,31] (as language develops, it is also important that the child uses language flexibly to develop connections and relationships among events and objects in their environment, and not to simply describe them); (c) early attention and social problem solving, with a focus on establishing contingencies that link action and movement [7,28]; and (d) responsive parenting, which represents strategies that support the development of skills in at-risk children [6,29]. Families with higher expectations for autonomy may be more likely to promote the flexible use of language, stronger attention, and independent movement early in development [29].

5.2. Later childhood

Many of the later developmental needs of the child with SBM involve school and learning. As a general principle, the approach to intervening in any area that involves school or behavior does not necessarily deviate because the child has SBM. Because there is little research specific to the learning needs of children with SBM, the working principles are that these children will benefit from interventions specific for their cognitive and academic difficulties, such as those for reading comprehension or math problem solving. These have been shown to be effective in other populations, such as children with learning or attention disorders [40], and which have, on a small scale, been shown to be effective for children with SBM [39]. One of the reasons that interventions for struggling students might be applicable is that many interventions are explicit in terms of identifying goals, providing external organizational structures to build skills in a step by step fashion ("scaffolding skills"), and teaching strategies directly by using associative learning to enable assembled processing.

5.3. Modal profile and individual variation

Understanding SBM requires that we identify the modal profile for outcome in a number of domains and then sculpt that outcome according to specific factors that we know produce individual variations in the profile. As we learn more about both group outcomes and individual function, we will be able to identify the best possible interventions based on information from both individual and group outcomes. The SBM profile, both modal and individual, is quite distinctive and is not captured simply by assigning these children to categories such as ADHD, nonverbal or right hemisphere learning disability, or a dysexecutive syndrome. Even though SBM shares some features with each of these conditions, and at a broad level such terms may facilitate communication around the modal profile, SBM is not well characterized by any of them. Assignment of any of these diagnostic labels in no manner dictates effective interventions. What is more important is accurately conceptualizing an individual's strengths and weaknesses in a way that enables them to receive services and to help guide the nature and content of such services.

5.4. Further research

Development of these guidelines also raised several questions that were not answered through a review of the existing published literature and which we identified as domains for further research. To what extent can

the early use of Magnetic Resonance Imaging (MRI) findings (e.g., malformations, dysplasia, reduced volume, and agenesis) predict domains of risk and identify potential early interventions to support development? What specific interventions in infancy are most successful in supporting the development of motor, cognitive, early literacy and numeracy skills? Multiple questions about shunt management have been raised, including those related to how the earliest time periods are managed in infants. The research has yet to detail the longterm effect of sequential monitoring of hydrocephalus on development. For example, is it better to shunt early and control hydrocephalus or to monitor ventricular expansion over time, and to identify the best indicators of the need for shunt diversion?

Also, there are still questions about how to manage some of the cognitive challenges that have been documented for many patients with Spina Bifida. One question is whether and to what extent different interventions used across the lifespan involving cognition, learning, and social skills are effective for persons living with Spina Bifida. Another is whether attention problems are best treated from pharmacological and/or non-pharmacological perspectives, as this has not been explored through randomized trials.

In addition, there are open questions with respect to the effects on neuropsychological function of recent changes in treatment for hydrocephalus, including prenatal repair in the Management of Myelomeningocele Study (MOMS) and the ETV/CPC procedures. There is a need for research to indicate how these treatments affect the health and physical and neurocognitive development of infants and children over time. The first report of school-age outcomes from the MOMS trial showed an improvement in motor functions and quality of life and a reduction of parental stress, but little evidence for improvement in cognitive functions and adaptive skills [42]. Furthermore, while many children who have undergone ETV/CPC procedures are still young, they may also demonstrate improvements over the more typical outcomes documented for those who were shunted in the past. Given increasing survival of individuals with SB into middle and later adulthood, the working group also noted a lack of information on the effects of SB and hydrocephalus on brain and neuropsychological functioning in middle and later adulthood.

The SB Guidelines for the Neuropsychological Care of People with Spina Bifida from the SBA's Fourth Edition of the Guidelines for the Care of People with Spina Bifida are based on significant advances in knowledge regarding both the modal neurocognitive profile of Spina Bifida as well as individual differences in that profile related to variability in neural structure, genetics, ethnicity, and the environment. Patterns of neuropsychological strengths and weaknesses that are evident in many individuals with Spina Bifida are discernable early in development and across the lifespan. Understanding the common neuropsychological profiles as well as individual differences in outcomes is critical for planning interventions at all ages and for tracking progress. Recognition of the specific aspects of the cognitive profile will also help medical providers to better understand how to work with their patients more successfully to facilitate their independent functioning over time. Despite considerable advances in understanding neuropsychological processes and outcomes in Spina Bifida, there is clearly need for further research on: 1) the effects of neurosurgical, pharmacological, neurobehavioral, and academic interventions on neuropsychological outcomes; and 2) the effects of Spina Bifida on the aging brain and ways to promote brain health in middle to later adulthood.

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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

The authors have no conflict of interest to report.

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