

Neuropsychology

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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 focused on patients who had hydrocephalus treated with surgical implantation of a shunt; however, the literature is just emerging on younger populations treated for hydrocephalus with different modalities, many of whom have similar cognitive profiles. It is important to identify this subgroup of patients with myelomeningocele, which makes up 90% of the population with Spina Bifida because individuals born with other types of Spina Bifida do not have these changes in neuroanatomical development 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 and vocabulary), and weaknesses when learning and performance involves the construction or integration of information (e.g. math problem-solving, reading comprehension). Many of these strengths and weaknesses are discernable across the lifespan.⁶⁻⁷

Motor Function: 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, which involves the relatively preserved basal ganglia,⁴ even though this type of learning may require more repetition and feedback than for typically-developing children.⁸⁻¹⁰

Perception: The ventral, object-based system involves detection of visual features and perception of categories (e.g., faces) and is critical for word reading. The dorsal, action-based system is responsible for the construction of visual representations and the linking of these representations to movement. This system relies on the posterior parietal region, which is disrupted by hydrocephalus. Children with SBM can identify faces and read words, but have difficulty with visual spatial reasoning and visually-guided goal-directed action.¹¹

Language: Strengths are noted in vocabulary and grammar. However, children with SBM experience challenges at the level of oral discourse, comprehension, and the use of language in context (pragmatics).¹² Individuals with SBM have difficulty in matching language output to a changing social language context.¹³⁻¹⁴ This has been linked to anatomic changes in the corpus callosum.¹⁵

Reading: Word recognition is often well developed¹⁶⁻¹⁷ reflecting compensation in middle temporal lobes.¹⁸ Difficulties in reading comprehension parallel those in oral language.

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 problem-solving skills.^{17 19-20} Problems with math, a long-term predictor of adult independence, are common in adults and children with SBM.^{2,21}

Attention: Many children with SBM meet criteria for Attention Deficit Disorder, Predominantly Inattentive Type (ADD).²²⁻²³ However, in contrast to children with ADD related to frontal lobe dysfunction, 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.²⁴⁻²⁶ Attention deficits involving posterior brain pathways may be a better way of understanding the self-regulation and organizational problems of individuals with SBM than is “executive dysfunction,” such as with traumatic brain injury and other injuries affecting frontal lobe function. Indeed, the frontal lobes are relatively spared in SBM. With sufficient repetition and error correction, people with SBM who have attention deficits can regulate their attention on specific tasks and learn content with persistence and over time.

Variability in the Typical Neuropsychological Profile: Understanding the variability in neuro-anatomic deficits, ethnicity, and the environment (socio-economic status and education) is the key to understanding individual (rather than group) differences in outcomes. Neurological status, including more severe hydrocephalus, repeated shunt malfunctions, and ethnicity predict poorer outcomes.⁵ Individuals with higher lesion levels have more severe neuro-anatomic brain malformations and higher rates of intellectual disability. Spinal defects at T12 and above are more frequent among individuals of Hispanic/Latino ethnicity. These populations also often have lower socio-economic status, diminished access to care and adverse outcomes attributable to social determinants of health.⁵

Psycho-educational testing provided by school districts identifies some, but not all, aspects of the SBM neurocognitive profile. A full neuropsychological assessment is recommended, when available, to document and monitor cognitive functioning in individuals with SBM.

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.

0-11 months

Clinical Questions

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 (e.g.,

malformations, dysplasia, reduced volume, and agenesis) to predict domains of risk and identify potential early interventions to support development?

Guidelines

1. Provide parents with formal teaching and intervention around the development of effective parenting practices for fostering developmentally appropriate and responsive parent-child interactions. 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).²⁷ (clinical consensus)
2. Closely monitor infants who have undergone prenatal treatment, given the paucity of literature on their long-term outcomes.²⁸
3. 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. Adaptive behavior assessments that are interview-based are easy to complete and sensitive to growth trajectories in development.⁷

1-2 years 11 months

Clinical Questions

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?

Guidelines

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 [INSERT EARLY INTERVENTION LINK]. 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)
2. Implement formal early intervention supports for language (delayed in onset, articulation difficulties, or unusual in pattern of development such as excessive imitation, difficulties in language comprehension), as well as physical and occupational therapy for independent mobility, strengthening, and functional activities that are essential for most children with SBM, along with parental involvement. (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 later in development.²⁷ (Family Functioning Guidelines)
4. Encourage the use of equipment that facilitates object exploration and manipulation because it can be essential to 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 [INSERT EARLY INTERVENTION LINK], occupational/physical therapy services, or from a physiatrist. (clinical

- consensus) (Mobility Guidelines)
5. Provide encouragement to participate in group learning experiences for children, 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 [INSERT EARLY INTERVENTION LINK]. (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. Monitor coordinated upper limb movement and attention multiple times per year in children with severe Chiari malformation, tectal beaking, and callosal hypogenesis.²

3-5 years 11 months

Clinical Questions

1. How does the relation 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?

Guidelines

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.²⁹ 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)
2. 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
3. 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.^{13,30} 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)
4. As part of the child's medical team, advocate for children to have 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)

5. Although it should not matter as to which of the 13 categories of special education a child is identified, “other health impaired” (or neurological disorder classifications in some states) helps 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.^{16,20-21}
6. Monitor development with assessments of early math and literacy skills to help establish more subtle difficulties with development³⁰ and the need for more tailored educational supports.
7. 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

Clinical Questions

1. How does the relation 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?

Guidelines

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 adjustment³¹ and adherence to dietary regimens. 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 clinical 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)
2. 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 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)

3. 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)
4. Carefully monitor children for the onset of attention problems, as they are often interpreted as motivational or behavioral issues and are often manifested as lack of focus, slow cognitive tempo, failure to initiate, and infrequently with hyperactivity or impulsivity.²³ Attention problems are correlated with the Chiari malformation, tectal beaking, and hypogenesis of the corpus callosum.^{24-26,30,32-33}
5. Follow American Academy of Pediatrics guidelines when evaluating for ADHD.³⁴ One-third of individuals meet criteria for ADHD, predominantly Inattentive Type on parent rating scales.⁵ Interventions for attention problems that involve medications may be tried, but clinical experience suggests that lower doses are effective and that many children with SBM do not respond robustly to stimulants³⁵, 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)
6. Monitor children 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.^{15,30} Over 25% of children with SBM have significant language and reading comprehension problems, which tend to be present both for listening and reading comprehension.⁵ 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.¹⁶
7. Monitor children for the development of math problems. Over 50% of children with SBM develop math difficulties.⁵ Assessment of mathematics should include assessment of complex calculation skills and, in the later grade school years, math word problems.^{19,20}
8. 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 children 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 children with math disabilities. Take advantage of children's strengths in rule-based learning by providing explicit, well-structured instruction.³⁶
9. 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)
10. 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)

13-17 years 11 months

Clinical Questions

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 all of 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?

Guidelines

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.^{11,19} Intervention programs should be maintained because the absence of intervention is associated with plateaus in skill development in most populations with disabilities.³⁶
2. Encourage participation in school-related and extracurricular activities and create vocational plans and transitional services with a focus on functional independence. (clinical consensus)
3. For students receiving special education services, the Individualized Education Plan [INSERT EARLY INTERVENTION LINK] (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 on 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)
4. Because social skills of individuals with SBM are strongly related to neuropsychological variables, namely attention and executive function,³⁸ consider using psycho-educational and/or neuropsychological assessments to inform psychosocial interventions and mental health supports.³⁹ (Mental Health Guidelines)
5. 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) Conduct yearly screening and timely referrals for appropriate diagnosis and treatment of anxiety and/or depression with psychotherapy and/or medication treatment as needed.³⁹ (Mental Health Guidelines, Quality of Life Guidelines)
6. 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 decision making 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 children 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.³⁴ (clinical consensus) (Bowel Function and Care Guidelines, Transition Guidelines, Urology Guidelines)

7. Advise children and/or their parents/guardians to obtain copies of psycho-educational and/or neuropsychological assessments. Explain that documentation of 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

18+ years

Clinical Questions

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?

Guidelines

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

- ensure the maintenance of learning skills essential for work and independence.^{21,24} Changes in these areas may be a sign of unidentified shunt failure or shunt dependency, or other significant medical problem requiring intervention. (clinical consensus). Full neuropsychological assessment is recommended for adults with SBM who experience cognitive decline and suspected shunt failure. (clinical consensus)
6. Monitor for mental health concerns and potential cognitive decline with aging. (Mental Health Guidelines)

Research Gaps

1. How can teams use early Magnetic Resonance Imaging (MRI) findings (e.g., malformations, dysplasia, reduced volume, and agenesis) to predict domains of risk and identify potential early interventions to support development?
2. What early interventions in infancy are appropriate for supporting the development of motor, cognitive, and early literacy and numeracy skills?
3. What is the long-term effect of sequential monitoring of hydrocephalus on development? Is it better to shunt early and control hydrocephalus or to monitor ventricular expansion over time? What are the best indicators of the need for shunt diversion?
4. How well do interventions used across the lifespan involving cognition, learning, and social skills work with persons living with Spina Bifida?
5. How are attention problems best treated from pharmacological and non-pharmacological perspectives?
6. 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?

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