Overview: Neuropsychological Functions in Spina Bifida

The neuropsychological outcome of spina bifida myelomeningocele (SBM) is a critical component of any comprehensive understanding of spina bifida (e.g., Copp et al., 2015) because of the focus on brain-behavior relations. Neuropsychological studies show a pattern of strengths and weaknesses involving motor, cognitive, academic, and adaptive functions (Dennis et al., 2006; 2010; Fletcher et al., 2008). In considering groups with SBM, a typical pattern is apparent, which we have termed the modal profile. In considering individuals with SBM, variability around the modal profile is apparent, but is principled.

The modal profile is apparent when the defining spinal lesion is a lower level myelomeningocele (below T1) with the characteristic Chiari II malformation and shunted hydrocephalus (SBM). It is not generally apparent in people with SBM and to upper level spinal lesions (>T1), economically disadvantaged children, and individuals with other spinal dysraphisms, who typically have normal brains (Fletcher et al., 2005). People with SBM have domain general difficulties in timing, attention, and movement (Figure 1) that are apparent and measurable in the first year of life (Lomax-Bream et al., 2007; Taylor et al., 2013). These domain general factors lead to domain specific problems in multiple content domains, generally with strengths and weaknesses within these domains (Figure 2). Because of the domain general factors associated with the spinal lesions, and congenital brain malformations involving the hindbrain, cerebellum, midbrain, and corpus callosum (Juranek & Salmon, 2010), individuals with SBM are stronger at retrieving information and generating material that has been associatively linked or stipulated, like vocabulary. This type of learning is rule-based or associative processing. Weaknesses involve learning that involves the construction or integration of a response, or assembled processing.

The modal profile in Figure 2 involves a range of functional outcomes that reflect dissociations between assets in associative processing and difficulties in assembled processing. Understanding neuropsychological strengths and weaknesses in SBM requires an understanding of processing demands, not content domains. For any content domain – motor, perceptual, language, reading,
and math – both assets and difficulties may be identified. Impairment within a content domain depends upon the extent to which a particular task draws upon associative versus assembled processing. These are relative strengths and weaknesses; an asset may not always represent completely normal performance. In addition, these patterns are developmental stable over time, but manifest differently depending on the developmental stage of the person. A general principle is that learning is facilitated when there is an opportunity to learn rules and procedures through repetition and error correction (Dennis et al., 2006).

**Motor**
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 (Dennis et al., 2010), and is an example of assembled processing. However, they can learn motor skills through repetition and correction of errors, which involves the basal ganglia, and is an example of associative processing. Lower levels of performance will be apparent on tasks that require repetitive motor actions, but the learning curve, which involves learning procedures and responding to error correction, is comparable to typically developing children (Colvin et al. 2003; Dennis et al., 2005; Edelstein et al., 2004). These patterns reflect the cerebellar impairment associated with the Chiari II malformation and the relative preservation of the basal ganglia in SBM (Juranek & Salmon, 2010).

**Perception**
Spatial perception in the brain involves two processing systems. The ventral, object-based system involves detection of features like contour, shape, size, and orientation and perception of categories such as faces. It is a critical system for word recognition. Because object and word processing are stipulated, this type of associative processing is preserved. The dorsal, action-based system is responsible for the construction of visual representations and the linking of these visual representations to movement, a form of assembled processing linked to the integrity of the posterior parietal region often disrupted by hydrocephalus. Children with SBM can identify faces and read words but have difficulty with visual relations and visually guided goal-directed action (Dennis et al., 2002).

**Language**
Within the language domain, strengths in vocabulary and grammar have been observed for many years, representing the learning of stipulated relations, or associative processing. Many individuals with SBM have significant problems at the level of narrative discourse that impairs comprehension and the use of language in context (pragmatics; Holck et al., 2009) linked to the integrity of the corpus callosum (Huber-Okrainec et al., 2005). Because this involves construction of meaning, it is an example of assembled processing. “Cocktail party speech” (Tew, 1979) in children with SBM represents difficulty in matching language output to a changing social language context.

**Reading**
Word recognition processes, which are stipulated relations, are often well developed in SBM, reflecting the adequate development of the phonological component of language because of preserved assembled processing (Barnes et al., 2001; 2007). In the brain this preservation reflects compensation and reorganization in middle temporal lobe regions associated with dorsal processing (Simos et al., 2011). Because of the same difficulties observed in comprehension of discourse, reading comprehension is often impaired even though the person may have access to the necessary vocabulary because of difficulty making inferences and to integrating background knowledge.

**Mathematics**
Many children with SBM are able to learn math facts and can retrieve information from the times tables. However, complex procedures that require multiple steps and algorithms are difficult. They are often poor at estimating quantities, with weak problem solving skills (Barnes et al., 2001; 2006;
English et al., 2009). Problems with math (and writing) are likely the most common problems experienced with adults and children with SBM and a long-term predictor of adult independence (Dennis & Barnes, 2002; Dennis et al., 2007).

**Attention**

Many children with SBM have significant problems with attention and meet criteria for ADHD (Predominantly Inattentive Type; Ammerman et al., 1998; Burmeister et al., 2005). The attention difficulties are not like those seen in children with developmental forms of ADHD, which involve problems with self-regulation and cognitive control. Rather, children with SBM are distractible, under-aroused, and overly persistent in controlling the focus of their attention. This type of attention is stimulus controlled and associated with a posterior attention system involving the midbrain and posterior cortex, and can be directly linked with problems alerting and oriented to external stimuli (Treble-Barna et al., 2014; Kulesz et al., 2015; Williams et al., 2013). With sufficient repetition and error correction, people with SBM can learn to regulate attention on specific tasks in order to learn content and are very persistent over time. Figure 3 shows performance on a task requiring sustained attention in children with SBM, ADHD, and typically developing controls (Brewer et al., 2003). Note that those with SBM perform as poorly as those with ADHD early in the task, but over time, perform more like typically developing children.

**Variability in the Modal Profile**

A child or adult with SBM who has a lower level lesion, Chiari II malformation, and shunted hydrocephalus will show the modal profile. But individuals with SBM vary in a number of dimensions, including environment, neurological status, and ethnicity. Understanding the variability in these dimensions of SBM the key to understanding individual (rather than group) differences in outcome. Variability in brain development will produce variations in how multiple areas of the brain operate in a coordinated fashion to effect associative and assembled processing. Environmental influences, including socio-economic status and education, also produce variations in the modal profile (Dennis et al., 2006). Neurological status, including more severe hydrocephalus, repeated shunt malfunctions, and ethnicity predict outcomes in SBM (Fletcher et al., 2005). The combination of upper level spinal defects, which reflect more severe brain malformations, and poverty, may lead to intellectual disabilities (Fletcher et al., 2005).

Understanding the timing, attention, and motor difficulties associated with SBM may yield a richer understanding of the neuropsychological strengths and weaknesses of children with SBM, and the bases for variation around the modal profile. The modal profile is not simply captured by single descriptors, such as nonverbal learning disability (ADHD, or dysexecutive syndrome. Although these depictions identify some pertinent features of the SBM modal profile, they do not capture its individual character, and they fail to address the principled sources of individual variability in the modal profile. In terms of executive functions, which are associated with SBM (Rose & Holmbeck, 2007; Tuminello, an attention model may be a better depiction of these problems with self-regulation and organization.

**Intervention**

Understanding the domain general basis for content domain assets and weaknesses may prevent
some of the negative outcomes in SBM. As a general set of principles, interventions should build on strengths in associative processing. Teaching explicitly with an understanding of the difficulties with construction and integration are key principles. Build on the strengths in vocabulary and rule-based learning, so that strategies involving verbal mediation are emphasized.

Fletcher et al. (2008) described in detail potential interventions for children with SBM in early childhood and in elementary school environments. In early development, difficulties in movement, timing, and attention control are apparent early in development and continue into adult life, interfere significantly with the development of the assembled processing and have a lesser impact on associative processing. Four areas that may be important to facilitating the early development of children with SBM include (a) early movement, with a focus on encouraging the child to initiate and respond to environmental contingencies that require action; (b) early language, to ensure that speech and vocabulary not become overdeveloped in the child with SBM at the expense of inference, context-sensitivity, and comprehension skills. As language develops, it is also important to help the child use language flexibly to develop connections and relations among events and objects in their environment, and not to simply describe them (Taylor et al., 2013); (c) early attention and social problem solving, with a focus on establishing contingencies that link action and movement (Landry et al., 2013); and (d) responsive parenting, which represents strategies that support the development of skills in at-risk children (Landry et al., . Families with higher expectations for autonomy may be more likely to facilitate the flexible use of language and stronger attention skills as well as to promote independent movement early in development (Landry et al., 2013).

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, which have been shown to be effective in other populations, such as children with learning or attention disorders (Fletcher et al., 2007; 2008). One of the reasons that interventions for struggling students might be applicable is that many are explicit in terms of identifying goals, scaffolding skills, and teaching strategies directly by using associative learning to enable assembled processing.

**Conclusions**

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 both individual and group outcomes. As we have seen, the SBM profile, 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 (Yeates et al., 2003), or a dysexecutive syndrome (Rose & Holmbeck, 2007; Tuminello. 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, it is well characterized by none of them, and assigning them to any of these diagnostic labels in no manner dictates effective interventions. What is more important is accurately conceptualizing their strengths and weaknesses in a way that enables them to receive services and to help guide the nature and content of such services.

**Overall Expected Outcomes**

- Optimal development of language, academic, and other learning skills.
- Optimal performance in school, university, and vocational settings.
- Acquisition of a job.
- Maximize participation in society.
- Maximize independence according to individual capabilities.
Functional Outcomes
- Utilization of learning skills is apparent in a variety of contexts.
- Enrollment in early pre-school intervention.
- Enrollment in appropriate educational environment.
- Participation in vocational assessment and training.

AGE RELATED RECOMMENDATIONS

Infancy
Infants with Spina Bifida are well-served by family centered programs oriented towards the development of motor, language, and cognitive skills. Parental participation is critical given the longer term impact of services that teach parents how to facilitate the child’s development through enhanced mobility and exploration of the environment, play, and communication activities.

• **Clinical Question:** What early interventions in infancy and the preschool years are appropriate for supporting the development of motor, cognitive, and early literacy and numeracy skills?

• **Guidelines:**
  - Child is enrolled in early intervention program prior to initial discharge in the hospital and receives appropriate related therapies: OT, ST, and PT. For children who are not in an early intervention program appropriate therapy can be ordered by primary care provider or spina bifida team providers.
  - Parents are receiving adequate information and supportive services.
  - Promotion of effective parenting practices for infant stimulation and care giving. Interventions fostering developmentally appropriate and responsive parent-child interactions have been shown to be effective for promoting early cognitive and social development in multiple high risk populations. Responsive parenting in infants and toddlers with spina bifida is associated with more positive cognitive and social language outcomes at 3 years of age and with better social problem solving at 7 years of age (Landry, Taylor, Swank, Barnes & Juranek, 2013). Therefore, interventions that foster responsive parenting are recommended for infants and toddlers with spina bifida (e.g., Play and Learning Strategies or PALS Program Landry, Smith, Swank, & Guttentag, 2008.)
  - Appropriate placement in family-based infant development programs as early as possible through state and public school (IDEA Part C, 619) resources.
  - Facilitation of parental participation in infant development programs, including the Individualized Family Service Plan (IFSP).
  - Monitor and evaluate onset and progression of physical, cognitive, communicative, and social development.

• **Clinical Questions:** How is the health and development of infants and toddlers changing with prenatal surgery (MoM trials)? How is the health and development of infants changing with the use of new surgical procedure such as Endoscopic Third Ventricleostomy/Choroid Plexus Cauterization (ETV/CPC) rather than shunting? How does monitoring for hydrocephalus and delayed shunting influence development?

• **Guidelines:**
  - For infants who have untreated hydrocephalus and developmental regression, or slower than expected development, shunt placement/ETV should be discussed with the neurosurgeon.
  - For infants repaired prenatally, close monitoring is indicated given paucity of literature on their long-term outcomes. (Adzick, 2011)
  - For infants who have not yet been surgically treated for hydrocephalus, but are monitored due to risk, assessment using infant development scales involving language, motor, and
social development is warranted, usually by a neuropsychologist, developmental pediatrician, or other developmentally trained professional to evaluate infants. Adaptive behavior assessments that are interview-based are easy to complete and sensitive to growth trajectories in development (Taylor et al., 2011).

- **Clinical Questions**: How can teams utilize early MRI findings (e.g., malformations, dysplasia, reduced volume, and agenesis) to predict domains of risk and identify potential early interventions to support development?

- **Guidelines**:
  - For infants who are in a high risk category, which includes those born prematurely secondary to prenatal surgery intervention and those with significant changes in brain development (above Chiari II malformation), the risk for early difficulties with coordinated upper limb motor movements and attentional focus is especially higher with tectal beaking and greater severity of the Chiari.

**Toddler**

In this age range, language functions can be delayed in onset or unusual in pattern. As with any child, intervention can be effective in stimulating and modifying language development. Because of lower limb paraplegia, movement may be restricted. Furthermore, difficulties in fine motor control can make object exploration difficult. Children with Spina Bifida need encouragement in moving about and exploring their environment (Taylor). Interventions oriented towards facilitation of motor development can be effective. Parents need assistance in developing appropriate expectations for the child and in engaging in interactions that facilitate language and motor development. Children of parents with higher expectations who facilitate attention, require movement, and support language development have better outcomes later in development (Landry).

- **Clinical Question**: What early interventions in toddler years are appropriate for supporting the development of motor, cognitive, and early literacy and numeracy skills?

- **Guidelines**:
  - Monitor and evaluate onset and progression of physical, cognitive, communicative, and social development.
  - Support parents in effective interactions with child that facilitate movement and exploration, language and communication, and play.
  - Equipment that facilitates object exploration and manipulation can be helpful. 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.
  - Early interventions for language difficulties if delayed in onset, characterized by articulation difficulties, or unusual in pattern of development (e.g. excessive imitation, difficulties in language comprehension). The use of sign language is recommended in conjunction with verbal speech therapy.
  - Physical and occupational therapy for independent mobility, strengthening, and functional activities with parental involvement.
  - 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; either through community groups or early intervention.

- **Clinical Questions**: How is the health and development of infants and toddlers changing with prenatal surgery (MoM trials)? How is the health and development of toddlers changing with the use of new surgical procedure such as Endoscopic Third Ventriculostomy/Choroid Plexus Cauterization (ETV/CPC) rather than shunting? How does monitoring for hydrocephalus and delayed shunting influence development?
- **Guidelines:**
  - Children of parents with higher expectations who facilitate attention, require movement, and support language development have better outcomes later in development (Landry).
  - Monitoring of hydrocephalus must be based on developmental assessments beyond determination of milestones, which are weak indicators of developmental difficulties. Periodic assessments with age appropriate assessments of early language skills can help identify more subtle difficulties with development. Ongoing monitoring of coordinated upper limb movement and attention is essential in children with greater severity of Chiari, tectal beaking, and callosal hypogenesis.

**Preschool**
Pre-school and transition planning are important in readiness for school placement.

In this age range, attention and self-regulation skills begin to emerge as a separate domain and are related to subsequent development of cognitive and social skills. Expectations for independent problem solving, responsibility, and social interactions become critical for school performance and psychosocial adjustment. Preschoolers with Spina Bifida show early manifestations of attention, pragmatic language, and math difficulties that subsequently emerge as major factors in academic and social adjustment (Barnes et al., 2011). Language comprehension problems may be overlooked because of well-developed vocabulary and conversational speech. Interventions may facilitate subsequent development of reading comprehension later in school (Fletcher et al., 2008). Monitoring of development with assessments of early math and literacy skills can help establish more subtle difficulties with development and the need for more aggressive early intervention programs.

- **Clinical Question:** How does the relation between the nervous system and mental functions among patients with Spina Bifida affect their learning in reading, mathematics, writing, social science and science, and how does it affect them at different developmental stages?
- **Guidelines:**
  - Children with more severe hydrocephalus, hypogenesis and/or severe hypoplasia of the corpus callosum and history of CNS infection are at greater risk for difficulties involving construction of meaning from language (Bradley et al., 2016; Huber-Okrainec et al., 2005).
  - It is important that teams advocate for children to enter formal educational programs through the public schools in order to have access to the scaffolding that can support the development of attention, self regulation, social interaction skills, and independent functioning skills. If parents choose another school setting then recommendations for support services and supplemental resources should be provided and accessed.

- **Clinical Question:** What do teachers, psychologists, and other professionals need to know about the development of people with Spina Bifida?
- **Guidelines:**
  - All individuals need to understand that if spina bifida is associated with myelomeningocele, it is not simply an orthopedic disorder (“physical disability”). The brain malformations and hydrocephalus (with or without shunting) affect learning, especially in areas that require the construction and integration of information, such as language or reading comprehension and mathematics (Barnes et al., 2001; 2007; English et al., 2009). Full neuropsychological evaluations that includes assessment of early literacy and numeracy skills can help to provide a more comprehensive understanding of strength and weakness profiles, as well as the significant discrepancies often present in their profiles. These findings can be related to neuroimaging findings for a more careful determination of the basis for any difficulties experience by the child.
  - It is important to advocate for eligibility for special education services under “Other Health Impaired” or other category that emphasizes the need for ongoing care and attention to
changes in brain development (e.g., “Neurological Impairment” in Massachusetts), not one isolated domain of concern. Many children with spina bifida are identified as “orthopedically impaired.” Although it should not matter in which of the 13 categories of special education a child is identified, “other health impaired” helps schools understand that potential learning difficulties are related to the underlying neurological disorder.

**School age**

Academic, attention, and behavioral difficulties are frequently manifested in school age children with Spina Bifida. These problems tend to be identified later in school, partly because of the early development of word recognition, rote numerical skills, and vocabulary skills (in children who are not socially and economically disadvantaged) that mask the presence of difficulties with math and reading comprehension in many children with Spina Bifida. Attention problems are often interpreted as motivational or behavioral and are often manifested as lack of focus, slow cognitive tempo, lethargy (failure to initiate), and infrequently with hyperactivity or impulsivity (Burmeister et al., 2003). All of these problems are linked to the impact of the brain anomalies associated with and unique to myelomeningocele, especially the severity of the Chiari, tectal beaking, and hypogenesis of the corpus callosum (Bradley et al., 2016; Kulesz et al., 2015; Treble-Barna et al., 2013; 2014a, b; Williams et al., 2013).

Each school age child with myelomeningocele needs at least one complete neuropsychological evaluation with an assessment of adaptive behavior early in schooling to identify strengths and weaknesses in cognitive development. Over 50% of children with myelomeningocele develop math difficulties; one-third meet criteria for ADHD-Predominantly inattentive type on parent rating scales; and over 25% have significant language and reading comprehension problems, which tend to be present both for listening and reading comprehension (Fletcher et al., 2005). Because of these common academic difficulties in spina bifida myelomeningocele, neuropsychological assessment needs to include assessment not only of word reading accuracy and fluency, but also of text-level reading comprehension (Barnes et al., 2007). Assessment of mathematics should include assessment of complex calculation skills and other aspects of mathematics in older children that become important for mathematical success by the end of the primary grades (e.g., math word problems; Barnes et al., 2006; English et al., 2009). Interventions for attention problems that involve medications can and should be tried, but clinical experience suggests that lower doses are effective and that many do not respond robustly to stimulants (Davidovich et al., 1999), 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 Question**: How does the relation between the nervous system and mental functions among patients with Spina Bifida affect their learning in reading, mathematics, writing, social science and science, and how does it affect them at different developmental stages?
- **Guidelines**:
  - Children with spina bifida myelomeningocele, particularly those with hydrocephalus or other changes in brain development, would benefit from a full neuropsychological assessment. Although school assessments can be helpful to track global intellectual and academic progression, they rarely assess the development of essential skills in attention, executive functioning, coordinated upper limb, and memory domains, as well as adaptive skill acquisition. Tectal beaking is directly related to the presence and severity of attention problems (Kulesz et al., 2015; Treble-Barna et al., 2014a; severity of hydrocephalus and corpus callosum malformations to integration of information and to construction of meaning from language (Bradley et al., 2016; Huber-Okrainec et al., 2005).
- **Clinical Question**: What interventions in school-age children support their cognitive development and academic achievement?
• **Guidelines**
  • Reinforce use of learning activities and technology in home.
  • Promote child’s participation in school related and extra-curricular activities.
  • If a child has a problem with reading or math, interventions like those used with children with learning disabilities are often effective. For example, although problems with word reading and phonological awareness are rare, when present treatment programs like those used with children with dyslexia are effective. Math problem solving interventions designed for children with math disabilities, for example, have been shown to be effective for adolescents with spina bifida myelomeningocele and math difficulties and take advantage of these children’s strengths in rule-based learning using explicit, well-structured instruction (Coughlin & Montague, 2011).
  • Writing programs need to be implemented using assistive devices as early as possible. Keyboarding is viable as an alternative to handwriting, but some practice with paper and pencil skills is useful. Keyboarding must be taught if it is to be useful. Accommodations for writing difficulties are critical components of the educational plan.
  • Bladder and bowel continence needs to be addressed and aggressively managed as improved continence translates to greater independence and social participation.
  • Evaluation and treatment for attention and behavioral difficulties, including didactic teaching of social skills if necessary. Evaluations for ADHD should follow American Academy of Pediatrics guidelines. (Fischer, Church, Lyons, & McPherson, 2015; Smith K, et al., 2016).

• **Clinical Question:** What do teachers, psychologists, and other professionals need to know about the development of individuals with Spina Bifida?

• **Guidelines:**
  • All professionals need to understand that an individual with myelomeningocele does not simply have an orthopedic disorder. The brain malformations and hydrocephalus (with or without shunting) affect learning, especially in areas that require the construction and integration of information, such as language or reading comprehension and math problem solving. Physicians and teachers working with child should be aware that 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 where the child’s participation at an early age facilitates independence and social adjustment. Abstract concepts about self-care typically are ineffective and not practiced. Careful attention to how dietary regiments are introduced-again as verbal, rule-based learning will cater to the strengths of many with spina bifida and result in greater independence and autonomy.

**Teenage**
The evidence indicates that students do complete high-school but do so at a lower rate than peers without Spina Bifida. Transition issues become paramount in the adolescent period. For adolescents with a disability who are served in special education, transition plans are required beginning at age 14 that specifically address the transition to the vocational and occupational domains.

• Participation in appropriate school, vocational, and other community resources maximizes student potential and entry into higher education and/or job market.
• Complexities of academic tasks requiring the ability to analyze, assimilate, and integrate information can be particularly difficult for adolescents with Spina Bifida.
• Early transition plans are essential for development of the capacity to assume the roles and responsibilities of the post high school environment and achievement of optimal independence.
• If an individual with myelomeningocele has not previously had a neuropsychological evaluation with an assessment of adaptive behavior it is imperative that one be obtained at this age prior to transition into the adult educational, vocational, and medical environments.
Clinical Question: How does the relationship between the nervous system and mental functions among patients with Spina Bifida affect their learning in reading, mathematics, writing, social science and science, and how does it affect them at different developmental stages?

Guidelines:
- Interventions addressing integration and assimilation of information with a specific focus on reading comprehension and mathematics problem solving. Intervention programs should be maintained because the absence of intervention is associated with plateaus in skill development in most populations with disabilities.
- Vocational and transitional services with a focus on independence.
- Promoting participation in school related and extracurricular activities.
- Obesity and bladder/bowel continence can be major issues affecting social adjustment and should be aggressively addressed. (Fischer, Church, Lyons, & McPherson, 2015)

Clinical Question: What interventions and programs in adolescence provide smoother transitions to post-secondary education and/or career/vocational training?

Guidelines:
- It is essential to have an established transition plan built into patient’s IEPs in order to ensure that appropriate referrals are made to adult agencies, appropriate life and vocational skill training occurs, and there are discussions about plans after high school.
- Social skills training may be useful given the tendency of many individuals with spina bifida to become withdrawn and to experience reduced social interactions as they enter adolescence. Behavior that was cute and charming as a child is not always conducive to friendships in adolescence.

Clinical Question: What do teachers, psychologists, and other professionals need to know about the development of people with Spina Bifida?

Guidelines:
- In addition to the cognitive and learning problems associated with the underlying neurological disorder, adolescents with spina bifida are vulnerable to feelings of depression secondary to reduced quantity and quality of social interactions. Structured opportunities for social interaction through school, church, and afterschool opportunities may be needed to decrease a potentially higher risk for depression. (Essner & Holmbeck, 2010)

Clinical Question: How do treatment teams help prepare all of their patients for the transition to adulthood, and the assumption of their own medical care? What indicators are helpful to team in identifying those who may require ongoing support for adequate management of their medical conditions?

Guidelines:
- If patients have not been fully evaluated, it is essential that a full neuropsychological assessment be completed prior to the child’s 18th birthday. These results insure that if a diagnosis of an intellectual disability is relevant, it occurs in a timely fashion.
- Full assessments are also essential to help identify cognitive strengths and weaknesses for those who are assuming responsibility for their own medical care. Although efforts should begin as early as possible, it may be necessary to work with the patient for several years to learn the skills necessary to understand and take responsibility for their own medical care. The transition to adolescence may present new challenges to this learning and regression may be present.

Adults

Individuals with spina bifida typically achieve levels of independence, academic completion, and employment that are lower than expectations based on their development of intellectual and literacy
skills (Dennis & Barnes, 2003; Dennis et al., 2007; Jenkinson et al., 2011; Stubberud et al., 2012).

- **Clinical Questions**: How do treatment teams help prepare all of their patients for the transition to adulthood, and the assumption of their own medical care? What indicators are helpful to team in identifying those who may require ongoing support for adequate management of their medical conditions?
- **Guidelines**
  - Vocational services addressing job skills, additional education, and related activities.
  - University-based ADA/504 services for people with disabilities.
  - Monitoring of cognitive skills, especially math (Dennis et al., 2007), memory (Treble-Barna et al., 2014b), and attention, to ensure maintenance of learning skills essential for work and independence. Changes may be a sign of unidentified shunt failure or shunt dependency.
  - Transition from parent focused interventions and communication strategies (e.g., phone calls to nurse) to methods that may be easier for the young adult to utilize consistently (e.g., online portals, email).
  - Skill building will be essential in order for young adults to shift to the adult care model where less coordination of services is often provided.

**RESEARCH GAPS**

1. How well do interventions used across the lifespan involving cognition, learning, and social skills work with the person with spina bifida?

2. How are attention problems best treated from pharmacological and non-pharmalogical perspectives?

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?

**TIPS**

How do familial, social or medical risk factors (e.g., socioeconomic status, genetic disorders, other medical complications, language delays), in addition to the expected Spina Bifida profile of gross and fine motor and attentional difficulties, reflect in a profile of cumulative risk? How can family systems with risk for slower cognitive development be supported?

- Patient who are born with a higher lesion level tend to also have more affected motor systems (not because they are linked, but because higher level lesions are also associated with more brain dysmorphology; Fletcher et al., 2005; Juranek & Salmon, 2010). Therefore, these patients require more intensive supports.

- Socioeconomic status may compound or be an additive effect on top of primary effects of SBM and largely affect growth in language and reading – because this profile for low SES children (i.e., higher nonverbal than verbal abilities) differs from what providers generally think of when they think of SBM (higher verbal and nonverbal abilities; Fletcher et al., 2005; Swartwout et al., 2010) it is essential that these skill developments are closely followed early in development.

- Language delays will affect the acquisition of reading including reading comprehension, social discourse, and mathematics.

At what age are neuropsychological assessments most beneficial and recommended?

- Early in schooling and in adolescence. The profiles are stable. More frequent developmental assessments are needed when they are younger.

How is inattention with SB related to typical ADHD?
• Inattention in people with myelomeningocele is well-established to emanate from the midbrain and tectum (Treble-Barna et al., 2014a; Kulesz et al., 2015), as well as the connectivity of these areas into parietal and frontal regions (Williams et al., 2015). As such, attention problems are quite different from those seen in ADHD and are manifested primarily in inconsistent attention focus, distractibility, excessive attentional focus (child is “stuck” on something), lethargy, and slow cognitive tempo and reduced alertness. The impulsivity and attention persistence problems characteristic of ADHD are less apparent (Ammerman et al., 1998; Burmeister et al., 2005).

Why do patients with SBM have a hard time getting started on tasks?
• It is essential for children with SB to be encouraged to be autonomous and independent as early as possible. If children are not, a more passive approach to everyday adaptation will emerge, especially in families that see the child as disabled and needing lots of help. Children function best when taught explicitly what to do through strategic knowledge and verbal procedures; this also helps to support generalization and transition.

How do we help parents and teachers understand executive dysfunction and how to train those skills?
• Children with SB need external structure, explicit guidance and strategies, and other approaches that take advantage of their strengths with rule-based learning. In the absence of evidence to the contrary, children with SBM who also have learning difficulties are likely to respond to specific interventions (e.g., in reading, in math etc.) that have been shown to be effective in individuals with learning difficulties and no SBM. Training executive functions as an isolated skill has not been shown to be effective (Jacob & Parkinson, 2015).

Citations


