DEVELOPMENTAL DISABILITIES
RESEARCH REVIEWS *16: 31–39 (2010)*

THE COGNITIVE PHENOTYPE OF SPINA BIFIDA MENINGOMYELOCELE

Maureen Dennis[1,2,3]* and Marcia A. Barnes[1,4]

[1]Program in Neurosciences and Mental Health, The Hospital for Sick Children, Toronto, Ontario, Canada

[2]Department of Surgery, University of Toronto, Toronto, Ontario, Canada

[3]Department of Psychology, University of Toronto, Toronto, Ontario, Canada

[4]Department of Pediatrics, University of Texas-Houston Science Center, Houston, Texas

A cognitive phenotype is a product of both assets and deficits that specifies what individuals with spina bifida meningomyelocele (SBM) can and cannot do and why they can or cannot do it. In this article, we review the cognitive phenotype of SBM and describe the processing assets and deficits that cut within and across content domains, sensory modality, and material, including studies from our laboratory and other investigations. We discuss some implications of the SBM cognitive phenotype for assessment, rehabilitation, and research.
Dev Disabil Res Rev 2010;16:31–39.

Key Words: spina bifida; cognition; academic skills

INTRODUCTION

A cognitive phenotype—a profile of mental and behavioral skills—is a product of both assets and deficits. Recent investigations have begun to specify what individuals with spina bifida meningomyelocele (SBM) can and cannot do, and why they can or cannot do it. Within a number of content domains, we have identified particular functions as either intact or impaired, generated hypotheses about underlying processing impairments that cut across content domains, and tested these hypotheses in neurocognitive experiments designed to challenge putative key processing deficits. Delineating the type of processing required for cognitive operations across different content domains, sensory modalities, and material types has helped to identify the characteristic cognitive-behavioral strengths and weaknesses associated with SBM and has allowed the neurocognitive profile of SBM to be linked in a principled fashion with the neurobiology of the disorder.

In this article, we review the cognitive phenotype of SBM and describe the processing assets and deficits, including studies from our laboratory and other investigations. We discuss some implications of the SBM cognitive phenotype for assessment, rehabilitation, and research. The overview in Figure 1 shows neurocognitive assets and deficits, both domain general core assets and deficits in timing, attention, and movement and domain-specific functional assets and deficits in perception, language, literacy, and numeracy. We argue that the cognitive phenotype in SBM is based on neurocognitive processing biases whereby some types of operations are intact and others impaired, rather than on either absolute or proportionate loss of function [Dennis et al., 2006a].

DOMAIN GENERAL CORE ASSETS AND DEFICITS

Timing

Timing and rhythm are essential components of movement and cognition [Ivry and Richardson, 2002]. Children with SBM have difficulties in the perception and production of timing and rhythm (Table 1). Perceptual timing deficits are revealed in elevated thresholds for discriminating brief (∼400 ms) temporal durations. Children with SBM have deficits in perceiving and producing rhythms. For rhythm production, a processing bias favors synchronization (responding in time to an externally paced rhythm) over entrainment (responding in time based on an internally generated model of the rhythm so as to produce the rhythm predictively).

Attention

For attention, a processing bias favors internally cued over externally cued attention, a component of which is the development of peripersonal spatial attention. Orienting to the external world and sense of peripersonal space are significantly impaired, even when internally cued attention works relatively well.

Attention includes both stimulus orienting and response control. Stimulus orienting is the automatic capture, disengagement, and shifting of attention to and from salient sensations [Posner and Peterson, 1990; Knudsen, 2007]. Response control is the voluntary selection of a motor response, a

Grant sponsor: Eunice Kennedy Shriver National Institute of Child Health and Human Development (NICHD); Grant numbers: P01-HD35946 and R01HD046609-04.
*Correspondence to: Maureen Dennis, Program in Neurosciences and Mental Health, Department of Psychology, The Hospital for Sick Children, 555 University Avenue, Toronto, ON, M5G 1X8, Canada. E-mail: maureen.dennis@sickkids.ca
Received 17 December 2009; Accepted 18 February 2010
Published online in Wiley InterScience (www.interscience.wiley.com).
DOI: 10.1002/mrdd.89

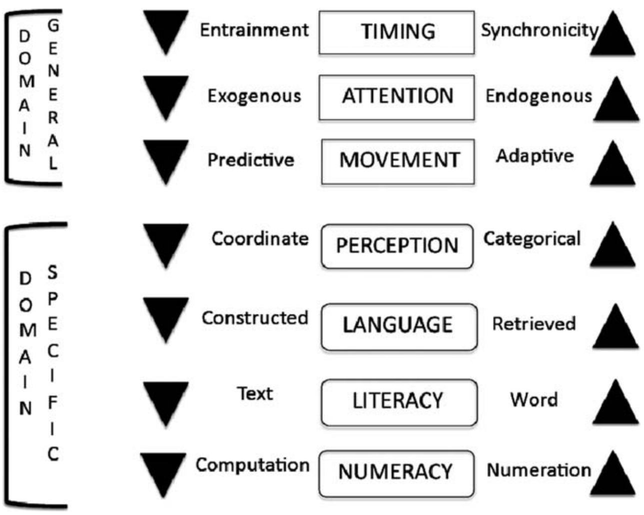

Fig. 1. Model of domain general and domain specific assets and deficits.

Table 1. Timing

Sensory motor timing
- ▼Perception of subsecond time intervals (∼400 ms) [Dennis et al., 2004]

Rhythm
- Synchronization
  - ▲Synchronized rhythm production [Dennis et al., 2004]
- Entrainment
  - ▼Entrained rhythm perception [Snow et al., 1994; Dennis et al., 2009a; Hopyan-Misakyan et al., 2009]
  - ▼Entrained rhythm production [Dennis et al., 2004]

For all tables, upward pointing triangles indicate that the function is intact, and inverted pointing triangles indicate that the function is impaired.

Table 2. Attention

Orienting
- To exogenous (external) cues
  - ▼Engage attention [Dennis et al., 2005a]
  - ▼Inhibition of return [Dennis et al., 2005b]
  - ▼Disengage attention [Dennis et al., 2005a]
- To endogenous (internal) cues
  - ▼Focus attention [Dennis et al., 2005a]
  - ▲Negative priming [Dennis et al., 2005a]

Peripersonal spatial attention
- ▼Attentional bias to left-right and inferior-superior hemispace [Dennis et al., 2005c]
- ▼Zone of peripersonal spatial uncertainty [Dennis et al., 2005c]

component of executive attention [Posner and Peterson, 1990; Rueda et al., 2005].

Children with SBM (Table 2) show relatively deficient stimulus orienting to exogenous (external, from the environment) cues; for instance, they require extra time to detach attention from an exogenous cue, resulting in an increased disengagement cost. Those with a characteristic brain abnormality of SBM, beaking of the midbrain tectum, show attenuated inhibition of return, expressed as a longer time to return to a previously attended cue location compared to a new cue location [Klein, 2000]. Exogenous orienting deficits are also apparent in infants and toddlers. Endogenous (internally cued) attention (e.g., negative priming, a longer time to attend to recent ignored stimuli; Tipper [1992]) appears relatively intact in SBM.

One aspect of spatial attention concerns peripersonal space, which is the part of egocentric space, within arm's reach, which is used for activities like picking up objects or drawing [Halligan et al., 2003]. Peripersonal space develops atypically in children with SBM, who differ from their age peers in terms of an exaggerated attentional bias to left hemispace, an abnormal attentional bias to inferior hemispace, and an enhanced Weber fraction, a larger zone of subjective uncertainty about peripersonal space.

Movement

Table 3 shows movement assets and deficits in SBM, in a group of studies using a range of effectors (eyes, hands, arms, and the speech articulatory mechanism). For movement, there is a processing bias in both preschoolers and school-aged children with SBM that favors motor adaptation and learning over predictive, dynamic motor control.

Domain General Core Deficits and Brain Abnormalities

The three core deficits, in timing, attention, and movement, we believe, are a direct consequence of the brain dysmorphologies of the Chiari 11 malformation [Raybaud and Miller, 2008; Juranek and Salman, in press] and associated hydrocephalus [Del Bigio, 2010]. In SBM, deficits in timing are related to the volume of the cerebellum [Dennis et al., 2004]; deficits in attention are related to the status of the midbrain, posterior cortex, and corpus callosum [Dennis et al., 2005a,b,c]; and deficits in movement are related to spinal cord dysfunction and cerebellar dysmorphologies that affect sensory-motor timing and motor regulation.

DOMAIN SPECIFIC FUNCTIONAL DEFICITS

Perception

Current perception models propose two different kinds of spatial relations between observers and objects [Kosslyn, 1987]. Categorical perception specifies discrete spatial relationships of visual primitives that may be described by categories (objects), feature groupings (faces), or verbal locatives (e.g., above, below, left, right); coordinate perception specifies precise spatial relations of visual primitives by means of coordinate metric values (e.g., "the line and the dot are 2 cm apart"). Categories and coordinates are basic computational elements for between- and within-category object recognition [Saneyoshi and Michimata, 2009]. In perceptual transformations (e.g., mental rotations of objects or reference frames) and multistable states (e.g., a picture with reversible figure and ground), encoding between observers and objects is vola-

Table 3. Movement

Eye movements
- ▲Visual fixation [Salman et al., 2009]
- ▲Vestibulo-ocular reflex in response to active head motion [Salman et al., 2008]
- ▲Saccade accuracy [Salman et al., 2005]
- ▼Smooth ocular pursuit generation [Salman et al., 2007]

Motor learning and adaptation
- ▲Adapting saccades to backward target displacement [Salman et al., 2006]
- ▲Adapting to prism-distorted visual input [Colvin et al., 2003]
- ▲Adapting drawing to mirror image [Edelstein et al., 2004]
- ▲Adapting ballistic arm movement to changes in relation between movement and vision [Dennis et al., 2006]
- ▲Learning manual rotation task [Wiedenbauer and Jansen-Osmann, 2007]

Ballistic movement
- ▲Ballistic arm movement [Dennis et al., 2006]

Dynamic motor regulation
- ▼Motor reaction time [Dennis et al., 2009b]
- ▼Motor speed [Zeiner et al., 1985; Ziviani et al., 1990]
- ▼Manual rotation time [Wiedenbauer and Jansen-Osmann 2007]
- ▼Limb taxis [Hetherington and Dennis, 1999; Lomax-Bream et al., 2007; Jewell et al., 2010]
- ▼Speech taxis [Huber-Okrainec et al., 2002]
- ▼Diadochokinesis [Jewell et al., 2010]
- ▼Eye–hand control [Wills 1993; Lomax-Bream et al., 2007]
- ▼Reaching [Norrlin et al., 2004]
- ▼Fine motor control and dexterity [Zeiner et al., 1985; Fletcher et al., 1995; Lomax-Bream et al., 2007]
- ▼Bimanual coordination [Hetherington and Dennis, 1999]
- ▼Drawing [Soare and Raimondi, 1977; Sandler et al., 1993]
- ▼Drawing a shape from an image in a mirror [Edelstein et al., 2004a]
- ▼Handwriting [Pearson et al., 1988; Ziviani et al., 1990; Barnes et al., 2004a]
- ▼Speech fluency [Dennis et al., 1987; Fletcher et al., 1995; Huber-Okrainec et al., 2002]

Table 4. Perception

Categorical
- Features
  - ▲Visual illusions of size, area, and length [Dennis et al., 2002]
  - ▲Auditory pitch category perception [Dennis et al., 2004]
  - ▲Auditory speech sound perception [Snow et al., 1994]
- Gestalts
  - ▲Forms [Fletcher et al., 1995]
  - ▲Figures [Hommet et al., 1999]
  - ▲Faces [Dennis et al., 2002]
  - ▼Fragmented objects [Dennis et al., 2002]
- Sequences
  - ▲Number-dot sequencing [Prigatano et al., 1983]
  - ▲Picture sequencing [Hommet et al., 1999]
- Relations
  - ▲Inferences about categorical spatial relations like up-down and left-right [Barnes et al., 2007]
  - ▲Virtual reality navigation based on spatial landmarks [Wiedenbauer and Jansen-Osmann, 2006]

Coordinate
- Metric
  - ▼Judgment of line orientation [Fletcher et al., 1995; Dennis et al., 2002]
  - ▼Geometry [Barnes et al., 2002]
  - ▼Spatial route finding [Dennis et al., 2002]
  - ▼Spatial route imaging [Dennis et al., 2002]
  - ▼Spatial route learning [Simms, 1987]
  - ▼Virtual reality navigation based on spatial coordinates [Wiedenbauer and Jansen-Osmann, 2006]
- Transformational
  - ▼Visual form constancy [Fletcher et al., 1995]
  - ▼Stereopsis [Dennis et al., 2002]
  - ▼Figure-ground perception [Fletcher et al., 1995; Dennis et al. 2002]
  - ▼Multistable visual illusions [Dennis et al., 2002]
  - ▼Egocentric mental rotations [Dennis et al., 2002]
  - ▼Extrapersonal mental rotations [Wiedenbauer and Jansen-Osmann, 2007]
  - ▼Mental rotation speed [Wiedenbauer and Jansen-Osmann, 2007]

tile. Categorical perception is intact in children with SBM, who identify features, gestalts, and relations based on categories or landmarks. Coordinate perception is impaired for illusory perception, pencil-and-paper, and virtual reality tasks. Table 4 shows perception assets and deficits in SBM.

Language

Language is a code by which meaning is expressed by linguistic representations [Dennis, 2009]. For language, a processing bias favors semantically retrieved over dynamically constructed meaning in SBM. Semantic information based on learned associations can be acquired, but language that must be constructed on-line through iterative cycles of activation, inhibition, and inferencing is impaired. Table 5 shows language assets and deficits.

Pragmatics is concerned with successful functional communication. One form of pragmatic communication is based on social and interpersonal principles like cooperation, turn taking, politeness, and irony; the other is based on textual rhetoric, including ease of processing, clarity, economy, and expressivity [Prideaux, 1991]. Interpersonal rhetoric is preserved in children with SBM, who are polite and friendly, sociable, cooperative, and interested in talking. In conversations, they initiate appropriate conversational turns and exchanges, using a mental state vocabulary. However, their textual rhetoric is impaired and their communication is difficult to process, uneconomic, and unclear.

Syntactic structures assign meaning [Caplan and Hildebrandt, 1988] of functional roles (e.g., who is acting, who is being acted on) and morphology attaches freestanding function words and inflectional morphemes in words and sentences. Phonology refers to the perception and integration [Plante et al., 2006] of features such as vowels, consonants, and syllables that have direct, identifying relationships with utterances. Children with SBM have generally intact syntax, morphology, and phonology.

Semantics is concerned with meaning, literal, idiomatic, and figurative. Semantics is variable in SBM. While semantic information can be activated to facilitate word finding, vocabulary development, and understanding of common idioms, there is impairment in the on-line, iterative cycles of updating and revision of meaning of text and discourse.

Literacy

Literacy involves coordinated skills that are, in part, developmentally staggered: mastering sound-letter correspondence; sight vocabulary; reading fluency; accessing word and sentence level meaning; and maintaining seman-

Table 5. Language

Pragmatics
- Interpersonal rhetoric
  - ▲Sociable, interested in conversations [Spain, 1974]
  - ▲Mental state vocabulary [Dennis et al., 1994]
  - ▲Uses mental state verbs to indicate perspective shifts [Dennis et al., 1994]
- Textual rhetoric
  - ▼Nonroutinized, on-topic, contextually and socially appropriate utterances [Spain, 1974; Culatta and Culatta, 1978; Culatta, 1980; Fletcher et al., 2002; Vachha and Adams, 2003]

Syntax
- ▲Syntactic development [Spain, 1974]
- ▲Syntax production [Byrne et al., 1990]
- ▲Syntactic complexity [Barnes and Dennis, 1998]
- ▲Syntactic comprehension [Dennis et al., 1987]

Morphology
- ▲Uses inflections and function words [Swisher and Pinsker, 1971]

Phonology
- ▲Rhyme-cued word finding [Dennis et al., 1987]

Semantics
- Literal meaning
  - ▲Vocabulary [Spain, 1974; Horn et al., 1985; Barnes and Dennis, 1998]
  - ▲Word finding to semantic cues [Dennis et al., 1987]
  - ▲Verb generation [Dennis et al., 2008a]
- Figurative meaning
  - ▲Common idiom comprehension [Huber-Okrainec et al., 2005]
- Text meaning
  - ▼Schematic structure [Dennis et al., 1994]
  - ▼Understanding of social scripts [Dennis and Barnes 1993]
  - ▼Coherence, clarity, economy, and plausibility of narratives [Dennis et al., 1994]
  - ▼Using context to understand idioms [Huber-Okrainec et al., 2005]
  - ▼Text-based inferences, integrating propositions across segments of text, and suppressing irrelevant semantic information [Barnes et al., 2004b]
  - ▼Making inferences that link knowledge and text [Barnes and Dennis, 1998]
  - ▼Maintaining mental models of situations in text [Barnes et al., 2007]

Table 6. Literacy

Phonological processing
- ▲Pseudoword reading accuracy [Barnes and Dennis, 1992]
- ▲Pseudoword reading fluency [Barnes et al., 2004b]
- ▼Phoneme deletion [Fletcher et al., 1995]

Decoding
- ▲Word reading accuracy [Barnes and Dennis, 1992]
- ▲Word reading fluency [Barnes et al., 2004b]
- ▼Noun reading fluency [Dennis et al., 2008b]

Text comprehension
- Memory based processes
  - ▲Activation of word meaning [Barnes et al., 2004b]
  - ▲Activation of character motivation and location [Barnes et al., 2007]
  - ▼Suppression of contextually irrelevant word meaning [Barnes et al., 2004b]
  - ▼Text-based and knowledge-based inference generation [Barnes and Dennis, 1998; Barnes et al., 2004b]
  - ▼On-line text revision processes [Barnes et al., 2007]
  - ▼Text-level fluency [English et al., 2010]
- Strategic processes
  - ▲Reading goal adjustment [English et al., 2010]
  - ▲Self-monitoring of comprehension [English et al., 2010]

tic coherence within and without the text through iterative comprehension cycles [Barnes et al., 2007]. Literacy also involves executive control whereby metacognitive goals affect strategic text processing [van den Broek et al., 2005].

For literacy in SBM, a processing bias favors word-level and some sentence-level processing over text-level operations that affects fluency and comprehension for texts. Table 6 shows literacy assets and deficits in SBM.

Children with SBM can read pseudowords, non-words that follow the rules of phonology, evidence of presumptive mastery of the basic rules for representing speech sounds visually. They read single words and have an adequate sight vocabulary. While they can rapidly access the names for written words and pseudowords, their text-level reading fluency is deficient.

For text comprehension, children with SBM activate a range of information within the written text and from semantic memory or world knowledge that facilitates word and sentence comprehension when revision and integration processes are not required. However, they fail to suppress contextually irrelevant meanings and are inefficient in making key inferences within text or between text and knowledge, showing difficulties in on-line iterative revision and integration. Executive control of text comprehension appears to be relatively intact in children with SBM, who can adjust the depth of their text processing to match higher order strategic goals and accurately judge how well they have understood what they have read.

Numeracy

Beginning in the preschool years with the acquisition of basic grouping, subitizing and counting skills, children gradually acquire the ability to perform operations on number, such as addition and division, and to apply number skills such as estimating, comparing, and problem solving [Mazzocco, 2009]. For numeracy in SBM, a processing bias favors procedural operations over de novo and relational processes that require the integration and application of mathematical information. Basic enumeration and calculation are acquired, albeit slowly, but estimation, problem solving, and mental calculation are impaired. Table 7 shows numeracy assets and deficits in SBM.

Enumeration skills are acquired in SBM, although development appears to be protracted. Preschoolers with SBM have difficulties with counting procedures, but school-aged children with SBM perform as well as peers on tests tapping knowledge of numbers such as reading numbers, understanding number series, fractions, and the like. Deficits in object-based addition and subtraction involving transformation on quantities are apparent in the preschool years. By school age, the data on calculation are mixed, with some studies suggesting that accuracy in both single digit and multidigit arithmetic may be a relative asset within the domain of mathematics, with proficiency by middle to late childhood in earlier learned and better practiced operations (e.g., addition, subtraction, multiplication versus division). In contrast, mental computations are deficient both at school age and in

Table 7. Numeracy

- Enumeration
  - Preschoolers
    - ▼Counting procedures [English et al., 2009]
  - School-aged children
    - ▲Numeration [Barnes et al., 2002]
    - ▲Rational numbers [Barnes et al., 2002]
- Calculation
  - Preschoolers
    - ▼Object-based addition and subtraction accuracy above subitizable range (sums or subtrahends 4 and above) [English et al., 2009]
  - School-aged children
    - ▲Single digit addition and subtraction accuracy [Barnes et al., 2006]
    - ▲Multidigit addition and multiplication accuracy [Barnes et al., 2002]
    - ▼Single digit addition speed [Barnes et al., 2006]
    - ▼Multidigit subtraction [Ayr et al. 2005; Barnes et al. 2006; but see, Barnes et al., 2002]
    - ▼Division [Barnes et al., 2002]
    - ▼Developmental maturity of calculation strategies for single digit addition [Barnes et al., 2006]
    - ▲Self-reporting of calculation strategies [Barnes et al., 2005]
  - Adults
    - ▼Computation accuracy and speed [Dennis and Barnes, 2002]
- Math applications
  - ▼Estimating [Barnes et al., 2002]
  - ▼Problem solving [Barnes et al., 2002]
  - ▼Adult functional numeracy [Dennis and Barnes, 2002]

adulthood. Math applications and problem solving based on manipulation of number and quantitative information are consistently impaired from childhood through young adulthood. Executive control of mathematics may be better than expected, and children with SBM can provide accurate reports of their own calculation strategies, suggesting that they have access to how they are solving mathematical problems even when their solution strategies are immature.

DISCUSSION

How can we characterize the SBM cognitive phenotype, outlined above? What are the real-world implications of the SBM cognitive phenotype for everyday function in individuals with SBM and for cognitive and educational rehabilitation of individuals with SBM? How does understanding cognition in SBM through experimental studies inform a research agenda for the future?

The SBM Cognitive Phenotype: Associative versus Assembled Processing

We have argued elsewhere [Dennis et al., 2006a] that the core of the processing bias in SBM concerns associative versus assembled processing. In SBM, associative processing is relatively intact, while assembled processing is relatively impaired. We do not suggest that processing differences within either individuals or groups with SBM are absolute, but rather, that they constitute systematic processing biases. Associative Processing is based on the formation of associations, enhancement, engagement, and categorization. It includes adaptive changes in response to stimulus repetition, as well as the activation and categorization of stimulus information. In individuals with SBM, strengths in associative processing facilitate temporal synchronicity, endogenous attention, adaptive movement, categorical perception, retrieved language, word-level literacy, and numeration and calculation procedures. Assembled Processing, in contrast, is based on on-line iterative cycles of activation, disengagement, and integration; it includes the creation of internal feed-forward models to guide performance over time. Weaknesses in assembled processing disrupt temporal entrainment, exogenous attention, predictive movement, coordinate perception, constructed language, text-level literacy, and most types of mathematical problem solving.

Implications of the SBM Cognitive Phenotype for Assessment and Rehabilitation

Delineating the SBM cognitive phenotype has several implications for assessment and rehabilitation of cognitive-academic difficulties in individuals with SBM. It promotes a more precise identification and classification of cognitive function; it delineates assets as well as deficits; it hones more global diagnoses to specific treatment plans, pointing the way to more SBM-targeted forms of cognitive and academic rehabilitation; and it focuses a research agenda for the future.

More Precise Identification and Classification of Cognitive Function

Individuals with SBM have functional assets in timing, attention, movement, perception, language, literacy, and numeracy, as well functional deficits in the same domains. It is misleading, therefore, to classify or diagnose by domain ("perceptual deficit," "motor deficit") because each domain has assets as well as deficits.

Individuals with SBM have functional assets in audition and vision, as well as functional deficits in the same sensory modalities. This means that assets and deficits cannot be classified according to sensory modality ("visual processing deficit"); the fact that the auditory modality has core deficits (in timing, above) and the visual modality has both assets and deficits in perception means that the cognitive phenotype of SBM cannot be explained by a simple dichotomy between intact auditory and deficient visual perception. Perceptual deficits in SBM do not involve inability to perceive wholes rather than parts or a generic problem in perceptual integration (children with SBM are generally able to perceive gestalt forms).

Children with SBM have apparently well-developed ordinality (sense of what comes first, second, etc) but poorly developed temporality (sense of how events occur in time). The term "temporal sequencing deficit" applied to a disorder is ambiguous, because it is unclear whether the problem is temporal or ordinal. Our data provide evidence for a functional separation of ordinality and temporality (see also [Ullén and Bengtssen, 2003], for a neural separation). Practically, children with SBM do not have a temporal sequencing problem but rather a problem in temporal motor regulation, which we believe is the basis of their functional difficulty with movement control, drawing, and handwriting.

Individuals with SBM have functional assets involving verbal and nonverbal content as well as functional deficits involving the same types of materials. Therefore, it is misleading to classify assets and deficits according to type of material (e.g., "non-verbal learning disability") because some non-verbal functions develop well in SBM and some verbal functions develop poorly; further, compared to those with SBM, children assessed as having a non-verbal learning disability show a different pattern of spatial perception dysfunction [Mammarella et al., 2009].

In short, while children with SBM have widespread cognitive and behavioral difficulties, these are not pervasive within a domain and do not involve one modality or one type of material. The SBM cognitive phenotype involves a complex pattern of cognitive function not well characterized by current dichotomies.

Delineating Both Assets and Deficits in SBM Cognitive-Academic Function

The cognitive phenotype of SBM involves both assets and deficits. Diagnostic evaluations and assessments often focus on the areas of deficit, yet experimental studies have identified assets in each content and academic domain. The more precise delineation of assets and deficits that is emerging from experimental studies of cognition in SBM is largely unexploited in the design of programs for motor, cognitive, and academic remediation. However, there is some preliminary evidence suggesting that tailoring interventions to these assets and deficits may be effective (e.g., for math) and that basing treatments on an incorrect and incomplete understanding of the core deficit may be ineffective (e.g., for attention).

On a wide variety of different tasks, and between two different conditions in the same virtual reality task, children with SBM can perform categorical but not coordinate perception tasks. That children with SBM have relatively good spatial orientation when they use landmarks provides an avenue for improving their extrapersonal orientation and ability to navigate through their external environment and community.

Clinical motor deficits are obvious in individuals with SBM; until recently, however, the extent of the relatively well-developed ability for motor adaptation and learning in eye, arm, and hand in SBM has been underestimated and has not formed an explicit component of programs to improve coordination and handwriting.

Crossdomain training is an underexplored area of rehabilitation in individuals with SBM. In children with SBM, training in physical rotations improves mental rotation skill [Wiedenbauer and Jansen-Osmann, 2007].

Honing More Global Diagnostic Groupings into Specific Treatment Plans

Attention

Approximately one-quarter of children with SBM have reported difficulties in attention [Colvin et al., 2003; Burmeister et al., 2005; Fletcher et al., 2005; Vachha and Adams 2005; Rose and Holmbeck, 2007]. Specifying the attention phenotype of SBM with experimental tasks has helped to understand how it overlaps with, and diverges from, the cognitive-behavioral phenotypes in other conditions. For example, individuals with SBM have difficulties with specific attention-orienting tasks, such as inhibition of return, that are performed well by those with Attention Deficit/Hyperactivity Disorder (ADHD) [Dennis et al., 2008b].

A better understanding of the attention phenotype in SBM helps make sense of some of the treatment outcome data. Children with SBM respond more poorly than children with ADHD to stimulant medication treatment [Davidovitch et al., 1999; Greenhill, 2002], suggesting that standard medication treatments for ADHD may be suboptimal for individuals with SBM, whose attention profile does not include the response control deficits that respond well to stimulant medication.

Executive function

Like many neurodevelopmental disorders, SBM is characterized by poor executive function on psychometric tests [Iddon et al., 2004; Rose and Holmbeck, 2007] and parent and self-reports [Mahone et al., 2002; Tarazi et al., 2008]. However, executive function deficits in SBM are not global. Individuals with SBM perform poorly on some but not all executive function measures [Brown et al., 2008]. Children with SBM do not make perseverative errors on the Wisconsin Card Sorting Test, and their poor performance on the Stroop task is due to slow naming speed [Fletcher et al., 1995]. Although children with SBM have difficulties disengaging attention [Dennis et al., 2005a], a key component of executive function, sustained attention, is relatively intact [Swartwout et al., 2008].

Children with SBM exhibit metacognitive control over their academic skills [English et al., 2010]. Like typically developing children, they take more time to read when the situation requires it (e.g., for study rather than for fun) and they are accurate judges of their own understanding. Metacognitive control may support academic remediation. In a case series of adolescents with SBM, Coughlin and Montague [in press] showed that a mathematics word problem intervention that involved learning and implementing executive strategies led to improved problem solving both postintervention and at long-term follow-up, as well as improvements in self-efficacy around math.

Executive function consists of representations, structured event complexes [Grafman, 2002] that are the basis of skills like metacognition and planning, and capacity-limited processing resources like working memory [Dennis, 2006]. The cognitive phenotype outlined here is consistent with studies showing that children with SBM have executive dysfunction, but, in SBM, executive representations may be more intact than executive processing resources, and representations like metacognition may be sufficiently functional to scaffold forms of cognitive-academic rehabilitation.

Some Issues for a Research Agenda for the Future

Crossdomain investigations

While it is clear that processing biases are related across core and functional domains, details of the relations remain to be specified to shape testable predictions about the nature of crossdomain associations.

Time, space, and number processing are complexly related [Cappelletti et al., 2009], although an association between peripersonal space and number is fairly well established. Numbers are conceptualized with a spatial metaphor (smaller numbers on the left and larger numbers on the right), and numerical information is represented spatially [for a review, see Umiltà et al., 2009], so common posterior parietal mechanisms may underlie the orientation of attention in physical space and along a mental number line. For instance, patients with right-sided neglect have a leftward bias when bisecting both physical lines and numbers [Pia et al., 2009]; the presentation of stimuli in near or far space modulates spatial attention for the mental number line [Longo and Lourenco, 2009]; and the direction of eye movements, left versus right, during arithmetic problem solving maps onto subtraction versus addition using symbols or objects [Knops et al., 2009]. Children with SBM have an exaggerated leftwards bias in peripersonal space; a demonstration that this is related to anomalies in their mental number line might enhance prediction of which children with SBM are most at risk for later math deficits.

Understanding SBM in the spectrum of neurodevelopmental disorders

One productive line of future research is to understand SBM in relation to other disorders, such as 22q11.2 deletion syndrome. SBM and 22q11.2 deletion syndrome have shared and unshared genes, brain, and cognition.

In SBM, the evidence for genetic anomalies concerns the folate and homocysteine pathways. Studying transmission disequilibrium of SNP alleles, Martinez et al. [2009] reported that anomalies in cystathionine-Beta-synthase, dihydrofolate reductase, methylenetetrahydrofolate reductase, and thymidylate synthetase conferred an increased susceptibility to spina bifida. Nickel et al. [1993] reported three patients with sacral or lumbosacral meningomyeloceles and congenital heart defects associated with deletion or microdeletion in the DiGeorge critical region (22q11) and a clinical diagnosis.

Both groups have reduced cerebellar volumes. In 22q12.1 deletion syndrome, Eliez et al. [2000] reported reduced cerebral and cerebellar volumes relative to controls, with vermal lobules VI–VII reduced in the midsagittal area [Eliez et al., 2001]. The scaling of cerebellum reduction in SBM is nonlinear. Juranek et al. [2010] found that while total cerebellar volume was significantly reduced in the SBM group relative to controls, after correcting for total cerebellum volume, and relative to controls, the posterior lobe was significantly reduced in SBM, the corpus medullare was not different, and the anterior lobe was significantly enlarged [see Juranek and Salman, 2010].

Cognitively, both disorders have significant problems in processing time, space, and number [Simon, 2008]. For 22q11.2 deletion syndrome, the underlying deficit may involve coarse granularity of processing involving reduced resolution of mental representations of spatial and temporal information [Simon, 2008]. We have argued in this article that the underlying deficit in SBM is a processing bias within and across domains according to which certain types of spatial and temporal processing are intact. Specific comparisons on the same neurocognitive tasks have yet to be made, so it is not at this point clear whether the two disorders differ in granularity of processing, type of processing, or both.

SUMMARY

Experimental investigations of the cognitive phenotype of SBM have been useful in providing a fuller and more nuanced description of cognitive assets and deficits. More generally, these investigations have provided a link to the observed clinical function, psychometric test performance, and academic profile of individuals with SBM. To be sure, much is yet to be learned about the SBM phenotype itself and the sources of variability in how it is expressed within the SBM population. Nevertheless, it provides a framework for ongoing empirical investigations. As a cognitive research agenda moves forward, we will have a better understanding of how to optimize assessment and intervention programs, and, in parallel, develop a fuller understanding of how SBM is positioned within the spectrum of neurodevelopmental disorders.

ACKNOWLEDGMENTS

The content is solely the responsibility of the authors and does not necessarily represent the official views of the NICHD or the National Institutes of Health.

REFERENCES

Ayr LK, Yeates KO, Enrile BG 2005. Arithmetic skills and their cognitive correlates in children with acquired and congenital brain disorder. J Int Neuropsychol Soc 11:24–262.

Barnes MA, Dennis M. 1992. Reading in children and adolescents after early-onset hydrocephalus and in normally developing age peers: comparison of phonological analysis, word recognition, word comprehension, and passage comprehension skill. J Pediatr Psychol 17:445–465.

Barnes MA, Dennis M. 1998. Discourse after early-onset hydrocephalus: core deficits in children of average intelligence. Brain Lang 61:309–334.

Barnes MA, Dennis M, Hetherington R. 2004a. Reading and writing skills in young adults with spina bifida and hydrocephalus. J Int Neuropsychol Soc 10:655–663.

Barnes MA, Faulkner H, Wilkinson M, et al. 2004b. Meaning construction and integration in children with hydrocephalus. Brain Lang 89:47–56.

Barnes MA, Huber J, Johnston AM, et al. 2007. A model of comprehension in spina bifida meningomyelocele: meaning activation, integration, and revision. J Int Neuropsychol Soc 13:854–864.

Barnes MA, Pengelly S, Dennis M, et al. 2002. Mathematics skills in good readers with hydrocephalus. J Int Neuropsychol Soc 8:72–82.

Barnes MA, Smith-Chant B, Landry SH. 2005. Number processing in neurodevelopment disorders: spina bifida myelomeningocele. In: Campbell JD, editor. Handbook of mathematical development. New York: Psychology Press. p 299–313.

Barnes MA, Wilkinson M, Khemani E, et al. 2006. Arithmetic processing in children with spina bifida: calculation accuracy, strategy use, and fact retrieval fluency. J Learn Disabil 39:174–187.

Brown TM, Ris MD, Beebe D, et al. 2008. Factors of biological risk and reserve associated with executive behaviours in children and adolescents with spina bifida myelomeningocele. Child Neuropsychol 14:118–134.

Burmeister R, Hannay HJ, Copeland K, et al. 2005. Attention problems and executive functions in children with spina bifida and hydrocephalus. Child Neuropsychol 11: 265–283.

Byrne K, Abbeduto L, Brooks P. 1990. The language of children with spina bifida and hydrocephalus: meeting task demands and mastering syntax. J Speech Hear Disord 55:118–123.

Caplan D, Hildebrandt N. 1988. Disorders of syntactic comprehension. Cambridge, MA: MIT Press.

Cappelletti M, Freeman ED, Cipolotti L. 2009. Dissociations and interactions between time, numerosity, and space processing. Neuropsychologia 47:2732–2748.

Colvin AN, Yeates KO, Enrile BG, et al. 2003. Motor adaptation in children with myelomeningocele: comparison to children with ADHD and healthy siblings. J Int Neuropsychol Soc 9:642–652.

Coughlin J, Montague M. The effects of cognitive strategy instruction on the mathematical problem solving of adolescents with spina bifida. J Spec Educ (in press).

Cullata B. 1980. Perceptual and linguistic performance of spina bifida-hydrocephalic children. Spina Bifida Ther 1:235–247.

Culatta B, Culatta R. 1978. Spina bifida children's non-communicative language: examples and identification guideline. Allied Health Behav Sci 1:22–30.

Davidovitch M, Manning-Courtney P, Hartmann LA, et al. 1999. The prevalence of attentional problems and the effect of methylphenidate in children with myelomenigocele. Pediatr Rehabil 3:29–35.

Del Bigio MR. 2010. Neuropathology and structural changes in hydrocephalus. Dev Disabil Res Rev 16:16–22.

Dennis M. 2006. Prefrontal cortex: typical and atypical development. In: Risberg J, Grafman J, editors. The frontal lobes: development, function and pathology. New York: Cambridge University Press. p 128–162.

Dennis M. 2009. Language disorders in children with central nervous system injury. J Clin Exp Neuropsychol; DOI: 10.1080/13803390903164355.

Dennis M, Barnes M. 1993. Oral discourse skills in children and adolescents after early-onset hydrocephalus: linguistic ambiguity, figurative language, speech acts, and script-based inferences. J Pediatr Psychol 18:639–652.

Dennis M, Barnes MA. 2002. Math and numeracy skills in young adults with spina bifida and hydrocephalus. Dev Neuropsychol 21:141–155.

Dennis M, Edelstein K, Copeland K, et al. 2005a. Covert orienting to exogenous and endogenous cues in children with spina bifida. Neuropsychologia 42:976–987.

Dennis M, Edelstein K, Copeland K, et al. 2005b. Space-based inhibition of return in children

with spina bifida. Neuropsychology 19: 456–465.
Dennis M, Edelstein K, Frederick J, et al. 2005c. Peripersonal spatial attention in children with spina bifida: associations between horizontal and vertical line bisection and congenital malformations of the corpus callosum, midbrain, and posterior cortex. Neuropsychologia 43:2000–2010.
Dennis M, Edelstein K, Hetherington R, et al. 2004. Neurobiology of perceptual and motor timing in children with spina bifida in relation to cerebellar volume. Brain 127:1293–1301.
Dennis M, Fletcher JM, Rogers T, et al. 2002. Object-based and action-based visual perception in children with spina bifida and hydrocephalus. J Int Neuropsychol Soc 8:95–106.
Dennis M, Hendrick EB, Hoffman HJ, et al. 1987. The language of hydrocephalic children and adolescents. J Clin Exp Neuropsychol 9:593–621.
Dennis M, Jacennik B, Barnes M. 1994. The content of narrative discourse in children and adolescents after early-onset hydrocephalus and in normally developing age peers. Brain Lang 46:129–165.
Dennis M, Jewell D., Edelstein K, et al. 2006b. Motor learning in children with spina bifida:intact learning and performance on a ballistic task. J Int Neuropsychol Soc 12:598–608.
Dennis M, Hopyan T, Juranek J et al. 2009a. Strong-meter and weak-meter rhythm identification in spina bifida meningomyelocele and volumetric parcellation of rhythm-relevant cerebellum regions. The neurosciences and music III: disorders and plasticity. New York Academy of Sciences 1169:84–88.
Dennis M, Jewell D, Hetherington R, et al. 2008a. Verb generation in children with spina bifida. J Int Neuropsychol Soc 14:181–191.
Dennis M, Landry SH, Barnes M, et al. 2006a. A model of neurocognitive function in spina bifida over the lifespan. J Int Neuropsychol Soc 12:285–296.
Dennis M, Salman MS, Jewell D, et al. 2009b. Upper limb motor function in young adults with spina bifida. Childs Nerv Syst 25: 1447–1453.
Dennis M, Sinopoli KJ, Fletcher JM, et al. 2008b. Puppets, robots, critics, and actors within a taxonomy of attention for developmental disorders. J Int Neuropsychol Soc 14:673–690.
Edelstein K, Dennis M, Copeland K, et al. 2004. Motor learning in children with spina bifida: dissociation between performance level and acquisition rate. J Int Neuropsychol Soc 10:877–887.
Eliez S, Schmitt JE, White CD, et al. 2000. Children and adolescents with Velocardiofacial Syndrome: a volumetric MRI study. Am J Psychiatry 157:409–415.
Eliez S, Schmitt JE, White CD, et al. 2001. A quantitative MRI study of posterior fossa development in velocardiofacial syndrome. Biol Psychiatry 49:540–546.
English L, Barnes MA, Fletcher JM, et al. 2010. Effects of reading goals on reading comprehension, reading rate, and allocation of working memory in children with spina bifida myelomeningocele and typically developing peers. J Int Neuropsychol Soc DOI 10.1017/S1355617710000123.
English L, Barnes MA, Taylor HB, et al. 2009. Mathematical development in spina bifida. Dev Dis Res Rev 15:28–34.
Fletcher JM, Barnes M, Dennis M. 2002. Language development in children with spina bifida. Semin Pediatr Neurol 9:201–208.
Fletcher JM, Brookshire BL, Bohan TP, et al. 1995. Early hydrocephalus. In Rourke BP, editor. Syndrome of nonverbal learning disabilities: neurodevelopmental manifestations. New York: Guilford Publications, p 206–238.
Fletcher JM, Copeland K, Frederick J, et al. 2005. Spinal lesion level in spina bifida meningomyelocele: a source of neural and cognitive heterogeneity. J Neurosurg 102:268–279.
Grafman J. 2002. The structured event complex and the human prefrontal cortex. In: Stuss DT, Knight RT, editors. Principles of frontal lobe function. New York, NY: Oxford University Press. p 209–235.
Greenhill LL. 2002. Stimulant medication treatment of children with attention deficit hyperactivity disorder. In Jensen PS, Cooper JR, editors. Attention deficit hyperactivity disorder: state of science best practices. Kingston, NJ: Civic Research Institute. p 9–27.
Halligan PW, Fink GR, Marshall JC, et al. 2003. Spatial cognition: evidence from visual neglect. Trends Cogn Sci 7:125–133.
Hetherington R, Dennis M. 1999. Motor function profile in children with early onset hydrocephalus. Dev Neuropsychol 15:25–51.
Hommet C, Billard C, Gillet P, et al. 1999. Neuropsychologic and adaptive functioning in adolescents and young adults shunted for congenital hydrocephalus. J Child Neurol 14:144–150.
Hopyan-Misakyan T, Schellenberg G, Dennis M. 2009. Perception of strong and weak meter rhythms in children with spina bifida meningomyelocele. J Int Neuropsychol Soc 15:521–528.
Horn DG, Lorch EP, Lorch RF, et al. 1985. Distractibility and vocabulary deficits in children with spina bifida and hydrocephalus. Dev Med Child Neurol 27:713–720.
Huber-Okrainec J, Blaser SE, Dennis M. 2005. Idiom comprehension deficits in relation to corpus callosum agenesis and hypoplasia in children with spina bifida myelomeningocele. Brain Lang 93:349–368.
Huber-Okrainec J, Dennis M, Brettschneider J, et al. 2002. Neuromotor speech deficits in children and adults with spina bifida and hydrocephalus. Brain Lang 80:592–602.
Iddon JL, Morgan DJR, Loveday C, et al. 2004. Neuropsychological profile of young adults with spina bifida with or without hydrocephalus. J Neurol Neurosurg Psychiatry 75:1112–1118.
Ivry RB, Richardson TC. 2002. Temporal control and coordination: the multiple timer model. Brain Cogn 48:117–132.
Jewell D, Fletcher JM, Mahy CEV, et al. 2010. Upper limb cerebellar motor function in children with spina bifida. Childs Nerv Syst 26:67–73.
Juranek J, Dennis M, Cirino PT, et al. 2010. The cerebellum in children with spina bifida and Chiari II malformation: quantitative volumetrics by region. The Cerebellum DOI 10.1007/s12311-010-0157-x.
Juranek J, Salman MS. Anomalous development of brain structure and function in spina bifida myelomeningocele. Dev Disabil Res Rev 16:23–30.
Klein RM. 2000. Inhibition of return. Trends Cogn Sci 4:138–147.
Knops A, Thirion B, Hubbard EM, et al. 2009. Recruitment of an area involved in eye movements during mental arithmetic. Science 324:1583–1585.
Knudsen EI. Fundamental components of attention. 2007. Annu Rev Neurosci 30:57–78.
Kosslyn SM. 1987. Seeing and imagining in the cerebral hemispheres: a computational approach. Psychol Rev 94:148–175.
Longo MR, Lourenco SF. 2009. Bisecting the mental number line in near and far space. Brain Cogn; doi:10.1016/j.bandc.2009.10.016.
Lomax-Bream LE, Barnes M, Copeland K, et al. 2007. The impact of spina bifida on development across the first 3 years. Dev Neuropsychol 31:1–20.
Mahone EM, Zabel TA, Levey E, et al. 2002. Parent and self-report ratings of executive function in adolescents with myelomeningocele and hydrocephalus. Child Neuropsychol 8: 258–270.
Mammarella IC, Meneghetti C, Pazzaglia F, et al 2009. Representation of survey and route spatial descriptions in children with nonverbal visuospatial) learning disabilities. Brain Cogn 71:173–179.
Martinez CA, Northrup H, Lin J-I, et al. 2009. Genetic association study of putative functional single nucleotide polymorphisms of genes in folate metabolism and spina bifida. Am J Obstet Gynecol 201:394.e1–e11.
Mazzocco MM. 2009. An introduction to the special issue: pathways to mathematical learning difficulties and disabilities. Dev Disabil Res Rev 15:1–3.
Nickel RE, Pillers DM, Merkens M, et al. 1993. Velo-cardio-facial and Di George syndromes with meningmyelocele and deletions of the 22q11 region. Eur J Pediatr Surg 3 (suppl 1): 27–28.
Norrlin S, Dahl M, Rösblad B. 2004. Control of reaching movements in children and young adults with myelomeningocele. Dev Med Child Neurol 46:28–33.
Pearson A, Carr K, Halliwell M. 1988. The handwriting of children with spina bifida. Zeitschrift für Kinderchirurgie 43:40–42.
Pia L., Corazzini LL, Folegatti A, et al., 2009. Mental number line disruption in a right-neglect patient after a left-hemisphere stroke. Brain Cogn 69:81–88.
Plante E, Holland SK, Schmithorst VJ. 2006. Prosodic processing by children: an fMRI study. Brain Lang 97:332–342.
Posner MI, Petersen SE. 1990. The attention system of the human brain. Ann Rev Neurosci 13:25–42.
Prideaux GD. 1991. Syntactic form and textual rhetoric: the cognitive basis for certain pragmatic principles. J Pragmatics 16:113–129.
Prigatano GP, Zeiner HK, Pollay M, et al. 1983. Neuropsychological functioning in children with shunted uncomplicated hydrocephalus. Child's Brain 10:112–120.
Raybaud C, Miller E. 2008. Radiological evaluation of myelomeningocele - Chiari II Malformation. In: Ozek M, Cinalli G, Maixner W, editors. Spina bifida: management and outcome. Milan: Springer. p 111–142.
Rose BM, Holmbeck GN. 2007. Attention and executive functions in adolescents with spina bifida. J Pediatr Psychol 32:983–994.
Rueda MR, Posner MI, Rothbart MK. 2005. The development of executive attention: contributions to the emergence of self-regulation. Dev Neuropsychol 28:573–594.
Salman MS, Dennis M, Sharpe JA. 2009. The cerebellar dysplasia of Chiari II malformation as revealed by eye movements. Can J Neurol Sci 36:713–724.

Salman MS, Sharpe JA, Eizenman M, et al. 2005. Saccades in children with Chiari type II malformation. Neurology 64:2098–2101.

Salman MS, Sharpe JA, Eizenman M, et al. 2006. Saccadic adaptation in Chiari Type II malformation. Can J Neurol Sci 33:372–378.

Salman MS, Sharpe JA, Lillakas L, et al. 2007. Smooth ocular pursuit in Chiari type II malformation. Dev Med Child Neurol 49: 289–293.

Salman MS, Sharpe JA, Lillakas L, et al. 2008. The vestibulo-ocular reflex during active head motion in Chiari II malformation. Can J Neurol Sci 35:495–500.

Sandler AD, Macias M, Brown TT. 1993. The drawings of children with spina bifida: developmental correlations and interpretations. Eur J Pediatr Surg 3 (Suppl 1):25–27.

Saneyoshi A, Michimata C. 2009. Lateralized effects of categorical and coordinate spatial processing of component parts on the recognition of 3D non-nameable objects. Brain Cogn 71:181–186.

Simms B. 1987. The route learning ability of young people with spina bifida and hydrocephalus and their able-bodied peers. Zeitschrift fur Kinderchirurgie, 42 (Suppl 1): 53–56.

Simon T. 2008. A new account of the neurocognitive foundations of impairments in space, time, and number processing in children with chromosome 22q11.2 deletion syndrome. Dev Dis Res Rep 14:52–58.

Snow JH, Prince M, Souheaver G, et al. 1994. Neuropsychological patterns of adolescents and young adults with spina bifida. Arch Clin Neuropsychol 9:277–287.

Soare PL, Raimondi AJ. 1977. Intellectual and perceptual motor characteristics of treated myelomeningocele children. Am J Dis Childhood 131:199–204.

Spain B. 1974. Verbal and performance ability in pre-school children with spina bifida. Dev Med Child Neurol 16:773–780.

Swartwout MD, Cirino PT, Hampson AW, et al. 2008. Sustained attention in children with two etiologies of early hydrocephalus. Neuropsychology 22:765–775.

Swisher LP, Pinsker EJ. 1971. The language characteristics of hyperverbal, hydrocephalic children. Dev Med Child Neurol 13:746–755.

Tarazi RA, Zabel A, Mahone EM. 2008. Age-related differences in executive function among children with spina bifida/hydrocephalus based on parent behavior ratings. Clin Neuropsychol 22:585–602.

Tipper SP. 1992. Selection for action: the role of inhibitory mechanisms. Curr Dir Psychol Sci 1:105–109.

Ullén F, Bengtsson SL. 2003. Independent processing of the temporal and ordinal structure of movement sequences. J Neurophysiol 90:3725–3735.

Umiltà C, Priftis K, Zorzi M. 2009. The spatial representation of numbers: evidence from neglect and pseudoneglect. Exp Brain Res 192:561–569.

Vachha B, Adams R. 2003. Language differences in young children with myelomeningocele and hydrocephalus. Pediatr Neurosurg 39: 184–189.

Vachha B, Adams R. 2005. Myelomeningocele, temperament patterns, and parental perceptions. Pediatrics 115:58–63.

van den Broek P, Rapp DN, Kendeou P. 2005. Integrating memory-based and constructionist processes in accounts of reading comprehension. Discourse Processes 39:299–316.

Wills KE. 1993. Neuropsychological functioning in children with spina bifida and/or hydrocephalus. J Clin Child Psychol 22:247–265.

Wiedenbauer G, Jansen-Osmann P. 2006. Spatial knowledge of children with spina bifida in a virtual language-scale space. Brain Cogn 62:120–127.

Weidenbauer G and Jansen-Osmann P. 2007. Mental rotation ability of children with spina bifida: what influence does manual rotation training have? Dev Neuropsychol 32:809–824.

Zeiner HK, Prigatano GP, Pollay M, et al. 1985. Ocular motility, visual acuity and dysfunction of neuropsychological impairment in children with shunted uncomplicated hydrocephalus. Childs Nerv Syst 1:115–122.

Ziviani J, Hayes A, Chant D. 1990. Handwriting: a perceptual-motor disturbance in children with myelomeningocele. Occup Ther J Res 10:12–26.