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Article *in* Journal of pediatric rehabilitation medicine · January 2008

Source: PubMed

NIH Public Access
Author Manuscript
J Pediatr Rehabil Med. Author manuscript; available in PMC 2010 August 25.

Published in final edited form as:
J Pediatr Rehabil Med. 2008 January 1; 1(4): 311–324.

Neurobehavioral outcomes in spina bifida: Processes versus outcomes

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Abstract

We review neurobehavioral outcomes and interventions for children with spina bifida. Focusing on children with spina bifida myelomeningocele, we contrast historical views of outcomes based on comparisons across content domains (e.g., language versus visual perceptual skills) with a view based on overarching processes that underlie strengths and weakness within content domains. Thus, we suggest that children with SBM have strengths when the skill involves the capacity to retrieve information from semantic memory and generate material that has been associatively linked or learned (*associative processing*) and general difficulties on tasks that require the construction or integration of a response (*assembled processing*). We use a hypothetical case to illustrate the differences in content domains versus general processes and also identify interventions that may be effective in addressing some of the cognitive and behavioral difficulties experienced variably by people with SBM. We extend these general principles to a discussion of variability in outcomes and use data from a large sample of children with spina bifida to illustrate the basis for this variability.

1. Introduction

The outcome of spina bifida, especially when the defining spinal lesion is a myelomeningocele with the characteristic Chiari II malformation and shunted hydrocephalus (SBM), involves a pattern of strengths and weaknesses motor, cognitive, academic, and adaptive functions [17]. When we speak of the neurobehavioral outcomes of SBM, we refer to groups and individuals. 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. Recent research has attempted to improve understanding of both groups and individuals by characterizing the basis for the variability around the modal profile. In this paper, we describe the modal profile, analyze the modal profile in terms of content domains vs. information processing, identify important sources of variability around the modal profile, and suggest targeted interventions based on the current evidence base. We ask:

1. What is the typical clinical presentation of motor, cognitive, academic, and adaptive behavior of children and adults with SBM, their *modal profile*?
2. Do the strengths and weaknesses in the modal profile represent differences between information content domains (between reading vs. math, for example, or between

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language vs. visual perception) or do they represent differences in how information is processed within and across information content domains?

3. Which factors within an individual with SBM lead to an atypical modal profile; that is, weakness in an area of typical strength or strength in an area of typical weakness?
4. Which evidence-based interventions can be helpful in addressing the typical cognitive and academic weaknesses of SBM?

2. Modal profile of SBM in a clinic setting

Consider the common scenario in which an individual with SBM returns to the clinic with new reports involving an assessment of cognitive and academic skills, completed by a school, and/or a battery of cognitive tests, completed by a neuropsychologist. The reason for initiating the school evaluation would have included concerns about special education eligibility and the need to develop an Individual Education Plan addressing the child's educational needs. The neuropsychological evaluation would have likely been initiated by a physician or parent in order to generate a comprehensive picture of the child's cognitive skills. In our experience, reports from either source often do not reflect awareness of the extensive research on cognitive and academic development in children with SBM; instead, reports often focus on the SBM motor deficits and attribute learning difficulties to motor, motivational, or behavioral problems. In this section, we review hypothetical but typical reports from a school psychologist and a neuropsychologist, and show how, in different ways, each provides evidence for a modal group profile in SBM.

2.1. A neurocognitive scenario

Jonny is a nine-year-old boy in grade 4 with SBM identified at birth. In addition to a repair of a lumbar-level level spinal lesion, he had a diversionary shunt implanted to treat his hydrocephalus, and, fortunately, has experienced no shunt difficulties that required neurosurgical treatment. Early CT scans revealed the characteristic Chiari II malformation, and an MRI scan at 4 year of age confirmed the classical Chiari II features associated with brain development in a small posterior fossa: beaking of the tectum, kinking of the medulla, and a small cerebellum. In addition, the corpus callosum was dysgenetic (the rostrum was missing) and hypoplastic (the posterior body was thinned).

In the past, Jonny was provided with occupational and physical therapies. At present, he has bladder and bowel incontinence, but is learning to catheterize himself. He is able to ambulate using crutches and braces, but also uses a wheel chair for longer distances. His middle-income parents are both supportive and actively involved in his care and education. As a fourth grader, he is eligible for special education as a child with orthopedic impairment, and, through special education services at school, has received 30 minutes per week of occupational and physical therapy focused on maintaining his capacity for ambulation.

Jonny's teachers find him something of an anomaly. Seemingly bright, communicative, and highly sociable, he had little difficulty learning to read words and recall basic math facts. Nevertheless, he has recently begun to struggle with math, especially long division, and one of his teachers commented that Jonny shows inconsistent understanding of what he reads and seems unable to integrate what he knows with the specific text in front of him.

Jonny's school completed an evaluation consisting of tests of intelligence and academic achievement, which is done with most students in the US suspected of a disability for which special education services are a possibility. Although there are many multifactorial IQ tests available, often with different theories of intelligence and psychometric approaches to their construction [32], the literature on intellectual outcomes in children with SBM has typically focused on discrepancies between scores on a verbal composite and a performance, nonverbal, or visual composite [43]. Tests like the Wechsler Intelligence Scale for Children-Revised (WISC-R) [41] predominate. Newer versions of these tests often provide more composites. For example, the fourth edition of the WISC generates composites for verbal comprehension, perceptual organization, processing reasoning, and working memory [42]. On this type of assessment, children with SBM would likely generate stronger performance on the verbal comprehension and working memory subtests than on the perceptual reasoning and processing speed subtests. The former composites assess skills like single word vocabulary and rote number repetition that are often strengths in children with SBM [19,43]. In contrast, children with SBM struggle with visual-perceptual integrative and organizational tasks, and timed paper and pencil measures of processing speed [20]. We will return to these issues below, but for the present let us note that the overall composite is not very meaningful when there are major discrepancies among these composites and our focus is simply on the well-identified contrast between stronger lexical and vocabulary skills (like those on any version of the WISC verbal composites) and difficulties with subtests assessing perceptual skills and speeded processing, typical composites of “nonverbal” IQ.

In this hypothetical report, Jonny's verbal comprehension composite score was in the average range, and his perceptual reasoning composite was in the low average range, some 10 points lower. The school psychologist also noted that Jonny is slow to complete timed parts of the IQ test, which were attributed to his motor difficulties. Teachers expressed concerns about Jonny's distractibility and difficulty initiating tasks and activities. On academic achievement tests, he could decode words at a high level and his sight word vocabulary and ability to sound out unfamiliar words were well above average, although his reading comprehension skills and math abilities were in the lower part of the average range, and significantly below his level of word decoding. The school psychologist noted that Jonny struggled on a test of reading comprehension that required him to read longer passages, even though he still had scores consistent with his overall IQ level. The psychologist reported that his math skills were inconsistent. Although he was successfully learning the multiplication tables, he had difficulty applying his knowledge of math facts to problems involving complex addition and subtraction, and double digit multiplication. His difficulty estimating quantities often produced answers that were not only incorrect, but were off by several orders of magnitude. The school committee convened to consider the report and other sources of evaluation data concluded that Jonny did not have learning needs because his achievement was consistent with his overall IQ score, which they told the parents they were required to use because of the state's special education guidelines. Nevertheless, the school committee did express concerns about how Jonny would fare on the state assessment to be administered in the spring, so they proposed attendance in preparation classes designed to assist him on this high-stakes assessment.

Jonny's parents also solicited an evaluation from the neuropsychologist associated with the clinic, who conducted additional tests of motor, spatial skill, language, attention, memory, and executive functions. In the motor domain, the neuropsychologist found that Jonny had severe bilateral fine motor difficulties, as well as difficulties copying geometric figures and judging the orientation of lines in two-dimensional space. In the language domain, he had well developed phonology, vocabulary, and grammar, but was not consistently able to understand by “reading between the lines” or by using the language context to derive meaning. Despite difficulties on a variety of tests of attention and executive functions, Jonny showed good effort and persistence on tasks in which he had to sustain attention over time. After a struggle to master the rules and strategies of some executive function tasks, he applied rules and strategies reliably after he had determined what they were, and he learned from his own errors to improve his performance. The neuropsychological report identified a number of strengths and weaknesses, and concluded that Jonny had a nonverbal learning disability, including an attention deficit disorder that required further investigation.

2.2. Understanding the modal profile

Each report is a meaningful, but separate, description of the same child. The reports are both organized according to domains, so that the school report focuses on intelligence and academic achievement, while the neuropsychological report addresses multiple motor and cognitive domains. Combining the information in the two reports produces a clear picture of a modal profile of SBM that is consistent with the literature [17,43,45], and that indicates (a) higher verbal than nonverbal IQ based on a traditional Wechsler or Stanford-Binet assessment; (b) better reading decoding than reading comprehension or math skills; (c) preservation of vocabulary and grammar, but poor language comprehension; (d) pervasive problems with fine motor, perceptual-motor, and spatial processing skills; (e) delimited difficulties on attention, memory, and executive functions; and (f) behavioral strengths involving social functions and persistence, but weaknesses in initiation and activation, cognitive flexibility, and self-regulation.

Having described the modal profile, we next attempt to explain it. Although the modal profile, especially as it is represented on IQ and achievement tests, is widely accepted in the clinical and research literature on SBM, an important issue is whether assets and deficits occur *between domains* or, alternatively, represent the operation of more general processes that cut *across domains*. If the former, all skills within a domain should be intact or deficient so that, for example, all language skills should be strengths and all visual perception skills should be deficits. If the latter, it is the process, not the content domain, that determines whether the skill will be intact or impaired. For example, inability to perform one type of cognitive processing will produce deficits in language and visual perception, while competence in another type of cognitive processing will produce language and visual perception strengths.

It has been difficult to settle this question because educational and neuropsychological research, like the two reports, is typically organized by content domains, not by cognitive processing. However, answering the question is important to clinicians working with SBM, not only to gain a deeper understanding of the ability profiles of these children, but also to generate meaningful interventions. If a child with SBM has domain-specific math problems, for example, then remediation can be math-generic. If a child with SBM has process-specific math problems, a significant portion of math-generic remediation (e.g., math fact retrieval) may be irrelevant while an adequate quantum of relevant remediation (e.g., in math procedures and problem solving) may not be offered.

2.3. What attributes are not measured by IQ tests?

The most commonly reported cognitive outcome for a child with SBM is a score on an IQ test. Many assume that IQ tests measure a child's general aptitude for school learning, representing a “general” intelligence factor (“g”) [33], and likelihood of responding to an intervention. However, composite IQ scores may not accurately depict a person's aptitude for learning. Some consider IQ to be a general attribute of a person [27], yet IQ scores are actually averages of many separate abilities, each of which reflect multiple genetic, neural, and environmental factors [10,13]. In SBM, there is no global outcome, as reflected in part by the commonly observed pattern of higher verbal IQ and lower nonverbal IQ. Global composites have lead to logical errors, such as the conclusion that early brain injury in humans is more benign than later brain injury [34], so neuropsychological research in SBM has moved towards identifying specific cognitive processes rather than composite IQ scores. Both IQ and specific cognitive measures are outcomes affected by early injury to the brain and the cascade of neural, experiential, educational, and environmental effects that follow.

If the goal is to find an overall average measure of abilities, IQ is a limited, but useful metric. To understand the discrepancy in verbal and nonverbal IQ, we must consider what IQ tests actually measure, especially in a child with an early brain injury; to understand why the discrepancy occurs, we must go beyond the observation that verbal IQ is higher than nonverbal IQ. To this end, we next consider two questions: Why is verbal IQ preserved? Why is nonverbal IQ lower than verbal IQ? The answers, we will show, can be found in an understanding the specific types of processes that are measured by the subtests of traditional verbal and nonverbal IQ measures.

2.4. Why is verbal IQ preserved?

Verbal skills on many IQ tests are assessed with subtests that involve vocabulary, verbal analogies (e.g., How are an apple and orange alike?), and verbal responses to everyday social situations (What should you do if in the movies you see a fire?). Although the surface content of these subtests is different, the underlying abilities that are assessed all involve access to the semantic and lexical content of language, typically organized under the rubric of “verbal comprehension” [32]. Measures involving mental arithmetic or working memory may also be included, but these verbal subtests predominate. The verbal subtests typically involve learning in which the meaning has been stipulated and learning occurs through association and repetition, with information retrieved from semantic memory. In many children with SBM, a type of associative learning is preserved, which we have termed *associative processing*, and which we have defined as the formation of associations, enhancement, engagement, and categorization through repetitive exposure with feedback from the environment. Associative processing underlies many of the strengths seen in individuals with SBM [17].

2.5. Why are nonverbal IQ scores impaired?

Depending on the subtests, the reduction in nonverbal IQ scores often observed in individuals with SBM, especially from WISC composites like perceptual reasoning and performance IQ, or the Stanford-Binet visual reasoning composite, can attributed to one or more of multiple factors. We will consider four of these factors: motor skills, synchronization of sensation and movement, spatial perception, and executive function.

Many nonverbal subtests place a premium on timed performance, so a person with fine motor deficits, characteristic of almost every child with SBM, will rarely obtain time bonuses. Fine motor deficits likely stem from the Chiari II malformation and its effects on the cerebellum and motor circuits. These deficits, however, are not simply deficits in motor coordination, but also the product of more general difficulties synchronizing sensation and movement. Many nonverbal tests, whether part of IQ tests or from other sources, involve skills such as using a pencil, rapidly underlining or copying symbols, or moving blocks into an array as quickly as possible. These skills require a person to be able to *precisely time* movements of the fingers and *synchronize* the response in real time based on feedback about the sensory consequences of motor events. Timing deficits are perceptual as well as motoric, and represent a fundamental deficit in the cerebellar circuit concerned with the synchronization of sensation and movement [16]. Thus, the reductions in performance IQ, perceptual reasoning, or even processing speed are not simply due to coordination of the fingers, but also involve complex functional deficits related to the cerebellar malformation and the effects of hydrocephalus on the brain.

Most nonverbal subtests require processing of spatial relations (e.g., constructing a puzzle or moving colored blocks so that they represent a pattern depicted on a two-dimensional card), which is a weakness in many with SBM [43], even without the demands for a timed motor response [21]. Nonverbal subtests do not often measure spatial functions that individuals with SBM can often perform at normative levels, such as face recognition, object matching, or shape matching.

Many nonverbal subtests also require an inferential, problem solving strategy that uses plans and schemata to assemble or construct a response that is novel in that it goes beyond the information provided in the stimuli, an example of an executive function. The parietal lobes, which are essential for this type of task, do not only match an external stimulus with an internalized representation, but also actively participate in the construction of the response and coordinate closely with systems that are regulatory and strategic that are mediated by the frontal lobes.

In sum, there are many reasons for lower scores on nonverbal test composites. Of itself, the finding of low nonverbal scores indicates little about the nature of cognitive impairments in SBM. As we shall see, conceptualizing performance in terms of the processes involved leads to more consistent and more interpretable results.

3. Associative vs. assembled processes

As these examples of why verbal IQ is preserved and nonverbal IQ is impaired demonstrate, individuals with SBM are able to retrieve information from semantic memory and generate material that has been associatively linked or learned, which we have termed *associative processing*. At the same time, they have general difficulties on tasks that require the construction or integration of a response, which we have termed, *assembled processing*. We define assembled processing as the construction and integration of information across various content domains [17].

The modal profile, illustrated in Table 1, involves a range of functional outcomes, and we argue that it reflects dissociations between assets in associative processing and deficits in assembled processing. The message of Table 1 is that predicting assets and deficits in SBM requires us to understand processing demands, not content domains. For any content domain – motor, perceptual, language, reading, and math – both assets and deficits may be identified. Impairment within a content domain depends upon the extent to which a particular task draws upon associative versus assembled processing. We now consider examples of associative and assembled processing within each domain in order to expand our discussion of the IQ discrepancy and to illustrate the general operation of associative versus assembled processes. It is important to note that these are *relative* strengths and weaknesses, so that a strength may not always entail completely normal performance.

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3.1. Motor

Children with SBM can learn motor skills when given the opportunity to do so by repetition and correction of errors. They have difficulty with controlled motor performance tasks that require adaptive matching of a motor response to changing visual information. Although they show low levels of performance on tasks that require repetitive motor actions, the slope of their improvement, which involves learning procedures and responding to error correction, is comparable to that of controls [11,18].

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3.2. Perception

Spatial perception involves two processing systems. The ventral, object-based system is supports detection of features like contour, shape, size, and orientation and perception of categories such as faces. The dorsal, action-based system is responsible for the representation of visual space in person-specific coordinates and the coupling of these coordinates to movement. Children with SBM can identify faces but have difficulty with visual relations and visually guided goal-directed action [15].

3.3. Language

Within the language domain, basic vocabulary and grammar develop well, but many individuals with SBM have significant problems at the level of discourse that impair comprehension and the appropriate use of language in context. While children with SBM can learn the meaning of an idiom and retrieve it from semantic memory, they have difficulty understanding an idiom whose meaning must be generated from the language context [26]. For example, our interpretation of “cocktail party speech” [36] in children with SBM is that it represents difficulty in matching language output to an evolving social language context.

3.4. Reading

Word recognition processes are often well developed in SBM, reflecting the adequate development of the phonological component of language. In contrast, comprehension of text is often impaired even though the child may know the words and meaning by sight. The basis for the impairment is likely the same as the impairment affecting discourse level comprehension [4], one involving failure to make inferences and to integrate world knowledge with ongoing context.

3.5. Mathematics

Children with SBM are able to learn math facts, such as those involved in retrieving information from the times tables. They have difficulty with algorithms,such as which operation, and in which order, to apply to number facts; they are poor at estimating answers and quantities; and their problem solving skills are often poorly developed [5].

3.6. Summary

In SBM groups, aspects of motor function, cognition, and learning that can be learned through association or repetition develop better than those that require integration of information, and this dissociation is the basis of the commonly observed modal profile. To be sure, associative and assembled processes are not directly observable or measurable, and must be instantiated in particular content domains.

Under typical conditions, then, a child with SBM will show the modal profile, including the discrepancy between verbal and nonverbal IQ, and better word recognition than mathematics skills. But not all individuals have the same history, and individuals with SBM vary in a number of dimensions, including biology, environment, neurological status, and ethnicity. Understanding the variability in these dimensions of SBM is the key to understanding individual (rather than group) differences in outcome.

Variability in brain development [23] 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. Neurological status and ethnicity predict outcomes in SBM [23]. We now consider how some atypical outcomes in SBM are related to biological and environmental variations, which is the basis for being able to predict outcomes not only in groups, but also in individuals.

4. Lack of discrepancy in verbal and nonverbal IQ

Table 2 presents means and standard deviations for verbal and nonverbal IQ proxies from the Stanford-Binet Intelligence Test – 4th edition (SB-IV) [38] and the word recognition (Basic Reading) and math computations (Calculations) from the Woodcock-Johnson Psychoeducational Test Battery – III [44]. The verbal IQ proxy is the vocabulary subtest from the SB-IV, while the nonverbal subtest is Pattern Analysis, a block construction task. We chose these two subtests because they are the best single indicators of the verbal and visual composites of SB-IV (as well as similar subtests on different versions of the WISC) [32]. In addition, we present results from adaptive behavior assessments of social communication and daily living skills from the Scales of Independent Behavior-Revised [7] to illustrate other strengths and weaknesses that affect activities of daily living. Social Communication assesses the quality of social interactions and relationships as well as rudimentary aspects of expressive and receptive language, which are known strengths in many with SBM [19]. Daily Living includes items related to toileting, dressing, other aspects of self-care, and independent living skills, which are known weaknesses in many with SBM, if for no other reason than the physical and orthopedic aspects of the disability [3].

We provide these data to illustrate the patterns and variations that occur within subgroups of children with SBM and have not conducted formal statistical analyses in this paper, although statistical evidence within the group with SBM can be found in other papers [23]. These data are derived from our sample of 322 children with spina bifida 7–17 years of age [24]. In this sample, 296 have SBM and 278 of these are shunted for hydrocephalus. Another 26 children have other spinal dysraphisms: two with shunted hydrocephalus, both with meningocles and no Chiari II malformation, with MRI evidence of aqueductal stenosis; three with ventricular dilation; 18 with meningoceles; and 3 with lipomas. These latter 21 children all had MRIs that were read as normal or other indicators of no brain abnormality if an older imaging modality (e.g., a CT scan) was used in the absence of a MRI scan.

In Table 2, data from 16 children with SBM are not included, representing two children with sensory disorders and 14 low-functioning children who were unable to perform the tasks. For the other 280 children with SBM, means and standard deviations are presented for three categories of outcome, with an average score of 100 and a standard deviation of 15. Note that for the entire sample of 280 children with SBM (Line 1 of Table 2), there is no major difference in verbal and nonverbal IQ. This lack of discrepancy, however, seems likely given the large and heterogeneous sample, as we show below. Higher scores on word recognition versus math, and social communication versus daily living, are clearly apparent, both consistent with the characterization of the child with SBM as sociable and able to read words, but weaker in math and struggling with activities of daily living. The latter weakness likely reflects the long-term impact of the spinal lesion on ambulation and movement, and the bladder and bowel difficulties.

In lines 2–4 of Table 2, scores are broken down by hydrocephalus status (shunted, arrested, or no hydrocephalus). Again, the discrepancy in verbal and nonverbal IQ is not apparent in the large group of shunted participants; in the small groups with arrested and no hydrocephalus, a pattern opposite of the modal pattern is evident for IQ, with lower verbal than nonverbal IQ. The differences among the groups are more apparent in the lower nonverbal IQ of the shunted group. In the academic and adaptive behavior domains, the differences reflect the much greater impairment in math and daily living skills in the shunted group, and the much larger discrepancy in word recognition vs. math, and social communication vs. daily living skills in the children shunted for SBM.

Table 2 then provides additional breakdowns of the group with shunted SBM, showing in lines 5–8 breakdowns according to lesion level, a biological variable. Here we also subdivide the group by ethnicity (Hispanic, non-Hispanic) since upper level spinal defects seem more common in Hispanic populations with SBM [23], where the heritability of lesion level has been reported to vary with ethnicity [40]. Here we see in line 5 the prototypical pattern for non-Hispanic children like Jonny with lower level lesions: lower nonverbal than verbal IQ, lower math computations than word recognition, and lower daily living than social communication skills. However, as line 6 shows, the discrepancy of verbal and nonverbal IQ is smaller for non-Hispanic children with *upper* level lesions, with a reduction in the overall level of performance in each domain. When we turn to the Hispanic participants in lines 7 and 8, we see the same effect of lesion level reducing overall performance in all three domains, with a similar pattern of performance in lines 7 and 8. However, comparing line 5 with 7, or 6 with 8, the verbal- nonverbal IQ discrepancy is *reversed*, showing lower verbal than nonverbal IQ for the Hispanic participants. Overall, such comparisons highlight that the modal profile is most apparent for the group of children who are not Hispanic and who have lower level (lumbar or sacral) spinal lesions.

The lesion level effect is not surprising. Children with thoracic level lesions not only have more severe movement disorders, but also have multiple indicators of more widespread brain dysmorphology [23]. One hypothesis about why Hispanic children show lower verbal than nonverbal IQ is that they are assessed in a minority language (English), which might explain their lower verbal IQ; however,22 of the children with shunted SBM were tested on comparable measures in Spanish, and they showed a similar pattern: verbal (M = 82; SD = 22); nonverbal (M = 87; SD = 15). Another hypothesis has to do with the large number of economically disadvantaged children in the Hispanic subgroup. Lines 9–12 show the groups subdivided by lesion level and socioeconomic status (advantaged,disadvantaged). As Table 2 shows, non-Hispanic and Hispanic children with SBM who are economically advantaged show a tendency for higher verbal than nonverbal IQ that tends to be larger in non-Hispanics; the opposite pattern is apparent for economically disadvantaged non-Hispanic and Hispanic children. In addition, there is an overall effect of SES, such that performance in all domains is lower in economically disadvantaged children. The discrepancy of word recognition vs. math, and social communication vs. daily living skills, is consistently evident in all subgroups.

Finally, consider the subgroup of 21 children with spina bifida who have other dysraphic lesions and no hydrocephalus (line 13, Table 2). All members of this subgroup had lower level spinal lesions, and the subgroups were roughly equal in ethnicity, with about 25% economically disadvantaged. Although the verbal IQ, word recognition, and social communication scores are somewhat comparable to the highest performing group of children with SBM and shunted hydrocephalus (line 5), their nonverbal IQ and math scores are much higher and in the average range. The impairment in daily living skills, while apparent, is much less evident than in other subgroups of children with shunted SBM.

Returning to the two evaluations of Jonny, we now have a more comprehensive understanding of how the academic and neuropsychological reports are both similar and different. Verbal IQ and word recognition are assets in children with SBM that reflect the operation of cognitive processes that are learned through association and repetition. The content varies, but the basis for the strength is a more fundamental cognitive process. Nonverbal IQ and math computation are deficits because they require integration of information. However, the model profile varies in a principled way, with sociodemographic factors (ethnicity), biological variables (lesion level), and environmental variables (socioeconomic status) reducing the level of performance and atypicality in the model profile. The operation of these factors is seen even in the adaptive behavior domain, which more directly reflects the impact of the orthopedic and urological complications of SBM, as well as difficulties with autonomy and independence as assessed by the items on the Daily Living Skills domain of the SIB-R.

Understanding the basis for content domain assets and weaknesses leads to general principles in interventions that help direct interventions that may prevent some of the negative outcomes in SBM. In addition, the link of associative versus assembled processes and variations in the physical phenotypes and the developmental environment of children with spina bifida illustrates that the problems experienced by Jonny are not just motivational, behavioral, or physical problems.

Of importance, the modal profile characteristic of SBM is not simply captured by single descriptors, such as nonverbal learning disability, attention deficit disorder, 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.

5. Interventions

Jonny will continue to experience problems in school. These are not new problems and their origins were much earlier than 9 years of age. In his situation, intervention will likely be delayed until he fails the state accountability tests, becomes depressed, or develops other problems that are secondary to his academic and social difficulties. Effective interventions need to start early in development and need to be content-specific, because teaching abstract skills in isolation may not generalize to the academic and behavioral domains.

5.1. Early development

The origins of the strengths and weaknesses of represent the modal profile can be seen in the early development of children with SBM. Deficits in movement, timing, and attention control, that are apparent early in development and continue into adult life [17], interfere significantly with the development of the assembled processing and have a lesser impact on associative processing. The implication for intervention in SBM is the need to think very early about the origin of deficits in assembled processing and to try and ameliorate them as much as possible through early interventions. Four areas that may be important to facilitating the early development of children with SBM include (a) early movement; (b) early language; (c) early attention; and (d) responsive parenting. Although there is little empirical evidence that such interventions are effective in spina bifida, there is substantial evidence from studies of other high risk populations [28].

5.1.1. Early movement—It is important to provide opportunities for infants and pre-schoolers with spina bifida to actively explore their environment because such experiences facilitate the development of mental representations and constructions of the external environment [6,9]. Although their difficulties with ambulation may interfere with these opportunities, it is relatively easy to conceptualize environments in which the child with SBM is allowed to struggle in order to promote opportunities for exploration and experience, in contrast to those that do not permit these opportunities and simply provide for the child's needs. In addition to physical therapy and occupational therapy, it is important to encourage parents to permit movement and not to simply solve movement problems for the child by always placing objects within their reach, and to move the child where they want to go. Although there is very little experimental research specific for children with SBM, there is evidence that early movement influences spatial cognition and the development of problem solving strategies [37].

5.1.2. Early language—Children with SBM have been described as showing an early onset of imitation skills [29]. As their language capabilities develop, it is common to observe good development of vocabulary and well-formed speech, which is also encouraging to parents and practitioners who care for the child. However, it is also important 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. Providing opportunities to elaborate on experiences with questions about the degree of coherence and connectiveness in the description is helpful. Early childhood intervention programs commonly provided in a school setting are often focused on producing well-formed speech and eliciting vocabulary. For children with SBM, these programs may provide insufficient focus on comprehension and pragmatic development, so additional interventions from speech and language therapists may be needed to foster the development of pragmatic language and inferencing. This becomes especially pertinent as the academic focus shifts beyond word reading to obtaining information via reading, with the volume increasing and the goal to understand what is read.

5.1.3. Parenting—Parenting of children with SBM is often atypical. Holmbeck and colleagues [25] found that parents of children with SBM, especially lower functioning children, tend to overprotect their child and exercise more control over their behavior, limiting opportunities for independence and autonomy. In contrast, allowing opportunities for choice, autonomy, and raising expectations may assist with improved outcomes.

Parent-based interventions may be an effective way to promote better outcomes in children with SBM. Landry et al. [28] described "responsive parenting," which represents strategies that support the development of skills in at-risk children. Families with higher expectations for autonomy may be more likely to facilitate the flexible use of language as well as to promote independent movement early in development. Responsive parenting involves: (a) the capacity for responding contingently to a child's signal as well as acceptance of the child; (b) emotional/affective support so that, for example, in a family experiencing distress, early counseling should be provided to prevent the development of maladaptive patterns of behavior; (c) language support, because providing rich language input that models an understanding of how and why things work and organize also helps with behavioral development.; and (d) attention support, because providing structure that helps a child to maintain his or her focus of attention has more effect on learning than simply re-directing the focus of attention. These attributes seem to support the development of attention and self-regulation skills, which facilitates the ability to address stress and novelty. The child learns explanations for feelings, more appropriate social understandings, and other related activities. In addition, the trust developed with parents permits opportunities to explore the environment and interact positively with the caregivers.

In summary, responsive parenting requires the ability to adapt to the changing needs of the child. In order to develop these kinds of parental behaviors, specific interventions may be needed to help parents understand how to engage in these behaviors as well as to reduce barriers for engaging in responsive parenting. Research on infants born prematurely with very low birth weight, who are at high risk for motor, attention, and language problems, shows that a focus on promoting autonomy may lead to enhanced cognitive development and improve psychosocial adjustment [28]; these authors have summarized a series of intervention studies that have been effective in facilitating cognitive and social skills through a focus on parent training. These interventions specifically target the kinds of cognitive processes we have described as assembled processes.

5.2. Later development

Many of the later developmental needs of the child with SBM involve school and learning. As we have noted earlier in this paper, certain skills seem to develop readily in children with SBM, while other skills emerge with effort and difficulty, often to surprise and consternation of parents and teachers. 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 principle is 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. 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 — in our terms, they use associative techniques to demystify and enable assembled processing. While there are other domains demanding intervention, we will discuss three that illustrate some general principles of intervention for children with SBM: attention and executive functions, reading and language comprehension, and math.

5.2.1. Attention—Many children with SBM show evidence for inattention and distractibility, with about one-third meeting research-base criteria for Attention Deficit Hyperactivity Disorder (usually the Inattentive Type) [2,8]. Patterns that involve impulsivity are relatively rare, but the inattention is associated with low arousal and sluggishness is common and is entirely consistent with what is known about the development of children with SBM. In addressing attention problems, there is little evidence that use of stimulants is specifically effective with children with SBM, and one of the few studies in the literature reported that attention deficits in children with SBM do not respond significantly to stimulant medication [12]. While these data do not mean that no children with SBM will benefit from stimulants, they do suggest that: a) stimulants will not be effective for many children with SBM; b) stimulants may be effective at lower doses than in children with primary or developmental ADHD; c) pre-medication evaluations should follow the guidelines established by the American Academy of Pediatrics [1]; d) medication trials should be conducted systematically, with well-operationalized criteria for responsiveness and preparedness to discontinue the medication or lower the dose if it is not effective; and e) careful attention should be paid to age of onset and pervasiveness criteria, as well as to the target symptoms, which typically involve inattention, disorganization, and poor self-regulation.

Children with attention problems perform well with teachers who are low-key, firm, and non-punitive, and in classrooms that are well organized and teacher-controlled. This means that the class should operate on a daily routine to which the teacher generally adheres on a regular basis, with the order of activities and amount of time devoted to various topics clearly displayed. The environment should be as quiet and visually neutral as possible. Additional modifications in the environment that are often useful include preferential seating near the teacher. Children with SBM who have attention problems should not be given complex instructions or be expected to follow directions on a single command. Instructions should be broken down and repeated on request. Teachers should use as much physical contact as possible as well as consistent attention-directing strategies (“listen to my voice”, “look at my nose”) to ensure adequate monitoring and attention skills. It may be helpful to teach children to assume a physical posture for attending, such as sitting at attention or in a “ready, set, go” position, to facilitate their ability to engage their attention skills.

Children with SBM require increased amount of structure, consistency, and help with organizational skills. They do not always know, or have access to, what they actually know. Many children with SBM will not be able to handle lists and should not be expected to take a list and complete the different activities on the list. They may have difficulty in situations which emphasize freedom of choice and which do not provide explicit structure or external support in completing tasks, so structure should be made explicit for the child. At home, the child should have a set time to do homework with frequent breaks. The parents should check each night to ensure that all work is completed. The child should use a backpack for school, which should be loaded the night before. Assignments should be kept in a divider notebook that is separated according to different subjects. Homework assignments should be placed in the appropriate subdivisions of the notebook. It would be helpful to make sure that the child's notebook has a zipper so that work will not fall out of it. When the child arrives at school, the first teacher each day should go through the notebook to ensure that the work is turned in to the appropriate teacher. The child should keep an assignment notebook or PDA where assignments are summarized by the teacher or another individual. When the child has to move from class to class, it is important to ensure that the child's locker is placed in a setting that is easy to reach and where his or her organizational deficiencies will not be a major obstacle. In general, helping children to know what they know, and to organize what they know, will support their executive skills.

When working with self-catherization and other daily living skills, helping the child with SBM focus their attention is important. The child may do best if the activities are verbally coded with ample opportunities for repetition and practice. Skills that are routine should be taught in ways that build upon associative processing. It may be very helpful to teach different activities involving attention and organization as part of exercises involving daily living and self- care skills.

5.2.2. Reading and language comprehension—For children with SBM, asynchrony in written language is the rule, and they may have a fluent sight vocabulary that is well in advance of what they understand. It is important to recognize that comprehension problems are evident in both oral and written language, and difficulties in comprehension have a similar basis in what the child hears and reads. This means that teaching the skills that promote comprehension will have a positive effect on both oral and written language comprehension. Key comprehension skills include: a) inferencing; b) activation of prior knowledge; c) comprehension monitoring; and d) sensitivity to structure of discourse or stories.

There are a number of effective interventions for facilitating comprehension skills. The key is to recognize that comprehension is not instantiated passively in language or in reading, but, rather, must be explicitly taught. While some children learn comprehension skills with minimal instruction, the skills themselves can be learned, and all children can improve their comprehension. Teaching comprehension skills involves the introduction of knowledge on what strategies are effective and when to employ them. For example, children with SBM can be taught strategies that help them attend to how a text or oral story is structured, how to summarize, elaborate, and connect different parts of stories and text, and how to monitor their own level of comprehension. Most children benefit from the explicit teaching and modeling of comprehension strategies in which the strategy is directly presented as a learning opportunity to the child. For example, the National Reading Panel Report [30] identified eight strategies for improved comprehension: comprehension monitoring, learning strategies and cooperative tutoring, graphic and semantic organizers, instruction in story structure, question answering, questioning generating, summarization, and multiple strategy teaching. Similarly, Vaughn and Klinger [39] found evidence that teaching strategies involving assistance in activating background knowledge, comprehension monitoring during and after reading, procedures using questioning, various methods that focus on the main idea in summarizing text and stories, explicit teaching of vocabulary, and graphic organizers all facilitated comprehension. Recent research on comprehension has highlighted the fact that it involves, not the passive reception, but the active assembly of meaning and that children with SBM are less able than their peers to assemble and construct meaning [4]. For the child with SBM, comprehension intervention involves explicit teaching of a variety of comprehension strategies and a focus on active, not, assembly of meaning of what is heard or read.

5.2.3. Math—In the modal math profile, children with SBM can generate and recall number facts, but have difficulties with estimating, using procedures such as carrying and borrowing, and solving math problems [5]. Some individuals with SBM may not have automatic access to math facts that they know, so will benefit from programs designed to enhance math fluency [5]. For those individuals with SBM who have profound difficulties with manipulating numbers in any form, interventions need to be focused more broadly on mastering math facts and on using numbers as part of adaptive, everyday functions, which, in this population, are an important correlate of perceived independence [14].

More generally, in teaching math to children with SBM, it is important to take advantage of their strengths in associative learning. They often benefit from math instruction in concrete, step-by-step approach where the math rules and steps are taught verbally as an entity to be memorized. Although this may seem somewhat antithetical to the idea of promoting assembled processing, it does take advantage of the strengths of individuals with SBM and seems more likely to facilitate math development. Thus, teaching math as a rule-based, step-by-step procedure with an emphasis on verbal mediation may be effective.

Word problems can be particularly difficult for children with SBM because they usually require the application of problem solving strategies. In general, it is helpful to explicitly demonstrate how to set up a word problem: using the text to identify missing information, constructing number sequences, and applying the correct calculations. Explicit, supportive instruction includes a component that addresses the transfer to other kinds of problems, given the general difficulties that children with SBM experience with novelty and forming connections across different classes of information [31].

6. 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, or a dysexecutive syndrome. Even though SBM shares some features with each of these conditions, and at a broad level such terms may facilitate communication around the modal profile, 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.

Although we have described only three school-based learning interventions, there is a wide range of academic interventions for children with problems in reading involving word recognition, fluency, and comprehension, written language difficulties including handwriting and story generation, and math involving fact retrieval and problem solving [22,35]. These interventions are often not applied in school settings, but have strong empirical bases in populations of children with developmental learning disabilities and ADHD. There are domains of potential interventions that we have not discussed in this paper, such as in the written language area.

One reason for focusing on strategy-based instruction is to emphasize the general theme of difficulty with any domain that requires the integration of information, or assembled processing. Whether this type of instruction will actually generalize across domains is not known, but would be interesting to study. In addition, the importance of explicit instruction for children with SBM in the strategies and procedures that promote assembled processing is also critically important. Many assume that children learn these skills best through a process of discovery and opportunities to independently construct knowledge. This is rarely effective for children with SBM (or for children with other forms of developmental disability). In general, children with disabilities require instruction that is more explicit and systematic in order to understand the constructs that are being taught.

In this paper, we have described the modal profile of children with SBM and targeted interventions that will help address their common weaknesses. However, we emphasize that variability is the norm for children with SBM and that averages do not necessarily apply to individual cases. The hypothetical example from the school report and neuropsychological evaluation are designed to try and link the evidence-base that has emerged on neuropsychological outcomes for children with SBM to children as they are seen in the clinic. However, individual cases always have primacy and there are no formulas that will work for every child with SBM. Each child and adult requires careful scrutiny in relation to the context in which development is occurring, and flexible, as well as intervention plans that change over time.

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Table 1

Strengths and weaknesses within cognitive domains for people with spina bifida myelomeningocele and hydrocephalus

Domain	Associative Processing Assets	Assembled Processing Deficits
Motor	Motor Adaptation	Motor Control
	Motor Learning	Visual-motor Tracking
Perception	Face Recognition	Spatial Relations
	Object Recognition	Mental Rotations
Language	Vocabulary	Making Inferences
	Grammar	Using Context
Reading Math	Word Decoding	Reading Comprehension
	Math Fact Retrieval (Times Tables)	Math Algorithms (Problem Solving)

Table 2

Patterns of strengths and weaknesses across content domains in subgroups of children with spina bifida myelomeningocele and other spina dysraphisms

Type	Subgroup	N	Verbal IQ	Nonverbal IQ	Word Recognition	Math Computations	Social Communication	Daily Living
1. SBM	ALL	280	86(19)	87(17)	92(25)	79(24)	89(18)	55(26)
2. SBM	H	258	86(19)	86(17)	92(25)	78(24)	88(18)	53(25)
3.	AH	17	88(13)	97(13)	96(23)	88(26)	99(16)	75(21)
4.	NH	5	91(23)	97(19)	102(6)	95(9)	98(7)	70(5)
5. SBM	NHsp,L	131	93(17)	87(18)	98(19)	81(22)	92(16)	60(23)
6.	NHsp,T	50	87(15)	84(17)	92(24)	76(22)	90(16)	46(23)
7.	Hsp,L	50	78(20)	89(13)	87(30)	82(27)	87(18)	51(26)
8.	Hsp,T	27	66(23)	77(12)	71(33)	60(24)	73(12)	40(28)
9.	NHsp,A	111	93(19)	89(17)	98(20)	84(22)	93(16)	59(25)
10.	NHsp,D	65	81(18)	85(15)	88(27)	77(26)	85(17)	54(25)
11.	Hsp,A	44	85(16)	83(18)	93(27)	78(22)	89(19)	46(24)
12.	Hsp,D	31	70(22)	77(11)	73(29)	60(22)	77(20)	43(24)
13.Other	ALL	21	91(20)	95(15)	103(25)	96(19)	94(14)	79(24)

SBM = spina bifida meningomyelocele; H = hydrocephalus; AH = arrested hydrocephalus; NH = no hydrocephalus; NHsp = non-Hispanic; Hsp = Hispanic; L = lumbar/sacral; T = thoracic; A = economically advantaged; D = economically disadvantaged; Other = other dysraphic lesions.