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MATHEMATICAL DEVELOPMENT IN SPINA BIFIDA

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Spina bifida (SB) is a neural tube defect diagnosed before or at birth that is associated with a high incidence of math disability often without co-occurring difficulties in reading. SB provides an interesting population within which to examine the development of mathematical abilities and disability across the lifespan and in relation to the deficits in visual-spatial processing that are also associated with the disorder. An overview of math and its cognitive correlates in preschoolers, school-age children and adults with SB is presented including the findings from a longitudinal study linking early executive functions in infancy to the development of later preschool and school age math skills. These findings are discussed in relation to socio-historical perspectives on math education and implications for intervention and directions for further research are presented.
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Spina bifida (SB) is a congenital neurodevelopmental disorder identified during gestation or at birth that is associated with high rates of math difficulties by school-age often in the context of adequate development of general cognitive abilities and word reading. As such, SB is a useful disorder for investigating how and why children develop problems with math and for studying some of the early developmental precursors of later emerging disabilities in mathematics. This article begins with an overview of SB: its epidemiology, pathophysiology, and a model of neurocognitive functioning that serves to organize findings across diverse cognitive outcomes including math. We then discuss ways in which this disorder has been used to understand mathematical ability and disability by using a life span approach that: (1) considers the natural course of mathematical abilities and their cognitive correlates in preschoolers, school-age children, and adults with SB; and (2) investigates potential developmental precursors of later developing mathematical skills using longitudinal methods. Possible implications for intervention and directions for future research are outlined.

SB: AN OVERVIEW

SB is a neural tube defect that results from complex gene-environment interactions. It is the most common disabling birth defect in North America, affecting the development of both spine and brain. SB occurs in ~0.3–0.5 of every 1,000 live births [Williams et al., 2005]. Atypical neuroembryogenesis affects the corpus callosum, midbrain and tectum, and cerebellum. Further complications arise from hydrocephalus, a buildup of cerebral spinal fluid in the brain that impedes myelination and damages gray matter, particularly in posterior brain regions. Shunting for hydrocephalus is required in many children with SB. The most severe form of this disorder is called SB myelomeningocele. It is characterized by a lesion though which the spinal cord protrudes, which can vary in location along the spine. The severity and nature of ambulatory and urinary complications vary depending on the level of this lesion. Higher level lesions are related to poorer outcomes, often resulting in difficulties with self-generated locomotion and bladder control, and also worse outcomes in some, but not all, aspects of cognition [Fletcher et al., 2004].

Neurodevelopmental outcomes across a number of domains for individuals with SBM have recently been linked to a small number of core deficits tied to the primary brain dysmorphologies of SBM that are evident from birth, persist throughout the lifespan, and result in a combination of spared and deficient processing within domains as diverse as motor function, perception, language, reading, and mathematics [Dennis et al., 2006]. Dennis et al. make a distinction between stipulated processing, which involves performance that is automatically activated and established through associations and repetition, and constructed processing, which relies on the integration of information from various sources and on-line adjustments of performance. The former type of processing is relatively intact in individuals with SBM, who show strengths in activation of stipulated representations including the ability to recognize faces, perceive objects from degraded visual cues [Dennis et al., 2002], retrieve small math facts (e.g., $2 + 3 = 5$) from memory [Barnes et al., 2006], and read words and access word meanings [Barnes and Dennis, 1992; Barnes et al., 2004b]. The latter type of processing is consistently deficient as seen in shifting between perceptual representations [Dennis et al., 2002], performance on larger sum computations whose answers are not reliably retrieved from semantic memory (e.g.,

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8 + 7) [Barnes et al., 2006], and integrating information within text to specify meaning [Barnes et al., 2007].

In the next section we present studies on math in children with SB that illustrate the ways in which SB has been used to: investigate the cognitive correlates and consequences of math difficulties from a life span perspective; test models of mathematical learning disability (MLD) subtypes; and study whether growth in particular cognitive abilities is related to mathematical outcomes in the preschool and early elementary school years.

LIFE SPAN STUDIES OF MATHEMATICAL PROCESSING IN SB: COGNITIVE CORRELATES, LONGITUDINAL PERSPECTIVES, AND LONG-TERM CONSEQUENCES

Infants and Toddlers with SB

The mathematical processing difficulties of individuals with SB are evident across the life span. As early as 36 months of age, preschoolers with SB have less well developed understanding of one to one correspondence, are less skilled at rote counting, and are less able to match on the basis of quantity than their typically developing peers [Barnes et al., 2005]. Fine motor and visual-spatial skills uniquely contribute to these emerging math abilities, with fine motor skill as measured by a task of fine motor dexterity (placing small grooved pegs into a pegboard) related to counting concepts and visual-spatial skill related to quantity comparison. These findings suggest that the difficulties in mathematical processing can be discerned in very young preschoolers with SB, certainly before the onset of any formal schooling, which has implications for early intervention and prevention. Moreover, the data also suggest that distinct mathematical abilities, even in the preschool years, may be supported by somewhat different cognitive and motor competencies. However, the developmental mechanisms by which deficits in fine motor skills and visual-spatial abilities might come to affect early math skills such as counting and quantity comparison have not been well-studied in this population or in the typically developing population. It has been proposed that in typically developing children across cultures there is a link between fingers and counting and simple arithmetic problem solving [Fayol et al., 1998; Butterworth, 1999; Noel, 2005], and also that pointing to and touching items may support the development of one-to-one correspondence knowledge [Alibali and DiRusso, 1999]. Deficits in fine motor skills and finger function and precision in upper limb control that are evident in infants and toddlers with SB [Lomax-Bream et al., 2007] could place early constraints on those aspects of counting and simple arithmetic that are supported by fluid and accurate finger and fine motor skills. Coupled with gross and fine motor restrictions, difficulties in visual attention (specifically in attention orientation involving attention to and disengagement from salient environmental stimuli) that are present from infancy [Taylor et al., in press] and that are related to midbrain dysmorphology [Dennis et al., 2005a,b] mean that very young children with SB do not efficiently explore space during a time of rapid development of visually guided behavior. Considering the importance of the coordination of visually guided motor skills such as reaching and exploration of the environment for cognitive development, early restrictions in these aspects of development could limit subsequent problem-solving and visual spatial capabilities [Thelen and Smith, 1994], skills that have been hypothesized to be important for mathematical cognition [Rourke, 1993; Assel et al., 2003]. These potential early developmental pathways to the evolution of difficulties in math in SB might be fruitful avenues for future longitudinal research investigations in both children with SB and in typical development. One example of such an approach is presented below.

> ***The mathematical processing difficulties of individuals with Spina bifida are evident across the life span. Math deficits in Spina bifida . . . are pervasive, persisting into adulthood and adversely impact quality of life.***

Longitudinal Study of Working Memory/Inhibitory Control in Infants and Mathematical Outcomes in Preschoolers and School-Age Children

Longitudinal investigations provide a unique opportunity for examining the cognitive underpinnings of later developing math abilities and impairments. A few recent longitudinal studies of math in typically developing children have focused on prediction of school-age math abilities from preschool cognitive processes. For example, these studies have investigated the relation of domain general abilities such as working memory and inhibitory processing [Blair and Razza, 2007; Mazzocco and Kover, 2007; Bull et al., 2008] or phonological processes [Hecht et al., 2001] and domain-specific abilities such as number sense [Locuniak and Jordan, 2008] in preschoolers to later math outcomes in kindergarten and the early primary grades. One of the advantages afforded by a neurodevelopmental disorder diagnosed at or before birth and associated with a high rate of school-age math disability, is that there is an opportunity to investigate growth in particular cognitive domains in infancy and relate them to later academic outcomes. The first study, to our knowledge, to examine the cognitive precursors of later developing math skills as early as infancy and to follow the same children through the preschool years and into the early school years, is a collaborative and multidisciplinary project involving the University of Texas Health Science Center at Houston and Toronto Hospital for Sick Children. English et al. [in press] evaluated whether infant executive functions predicted an array of preschool and school-age math skills. Although there is no consensus regarding an accurate definition of executive functions, many models highlight the central importance of both working memory and inhibitory control. Working memory is the ability to hold information on-line while engaging in simultaneous mental processes. Inhibitory control is the ability to suppress prepotent responses in favor of subdominant responses. A delayed response task, purported to measure these two constructs [Diamond and Doar, 1989], was administered to infants three times between 12 and 26 months. The nature of this task makes it difficult to discern the unique contributions of working memory and inhibitory control. However, failures of working memory/inhibitory control are inferred when the child searches under a cup that the reward (in this case, a Cheerio) was hidden under in the previous trial even though they subsequently saw the examiner place the reward under the other cup.

Latent growth curve modeling (LGM) was used to assess change in infant executive functions and to determine whether initial level and growth in these early executive functions predicted later developing preschool and school-age math skills. LGM is a flexible technique that offers various benefits compared to more traditional statistical techniques such as repeated-measures ANOVA. LGM can retain missing data by computing maximum likelihood estimates. LGM can also simultaneously evaluate growth in a construct as a dependent variable and as a predictor of subsequent outcomes. Two change parameters are evaluated in a growth curve model: intercept and slope. Intercept is the mean performance on a task and slope is the mean rate of change in a construct over time.

In general, working memory/inhibitory control predicted performance on a range of informal math outcomes at 60 months of age such as counting and object-based addition and subtraction [based on Jordan et al., 1992]. Performance on standardized tests of single and multidigit arithmetic at 7.5 years of age was also predicted by level and growth in infant working memory/inhibitory control. Interestingly, comparable trends were not as robustly observed for reading outcomes, particularly not at school-age. These different patterns suggest that there may be discrepant pathways to math and reading development. It is possible that reading places only developmentally limited demands on the executive system. Unlike math, reading becomes less effortful and more automatic over time; although some aspects of math (e.g., access to math facts and procedures in multidigit arithmetic) may become more fluent with experience, many aspects of math require new learning [LeFevre, 2000]. Therefore, reading may only engage the executive system when knowledge is being acquired and may also figure in the performance of older children with reading disabilities, who have often not automatized basic sound symbol correspondences. In contrast, math becomes increasingly complex as children get older and places considerably higher demands on executive processes.

The results from our longitudinal study on SB and learning suggest that specific cognitive processes, namely working memory and inhibition, may be instrumental for the development of math proficiencies. These findings are consistent with those from other recent studies and with some views on the relation between executive skills and math. It has been found that working memory is related to certain types of mathematical problem solving skills in typically developing preschoolers [Bisanz et al., 2005; Klein and Bisanz, 2000]. Blair and Razza [2007] found a similarly strong relation between measures of inhibitory control, effortful control, and math knowledge in a group of high-risk children attending kindergarten. Working memory and inhibitory control have also been found to predict performance on math word problems [Espy et al., 2004]. Children with severe MLD have difficulties with cognitive tasks that tap working memory and some aspects of their mathematical performance are either fully or partially mediated by working memory [e.g., Geary et al., 2007].

Results from our longitudinal investigation support hypotheses about the importance of certain cognitive systems such as working memory/inhibitory control in the development of math abilities, particularly when contrasted with the development of reading...

Domain-general theories of mathematical development and disability stipulate that certain core cognitive systems are foundationally involved in the emergence of math competencies. These systems include: the central executive, which involves attentional and inhibitory capacities; the language system, which is important for representing and manipulating mathematical knowledge for storage and retrieval; and the visuospatial system, which is responsible for understanding visual and spatial information [Geary et al., 2007]. In contrast to more domain-general theories of math development, domain-specific explanations stipulate that a specific number-processing deficit underscores math difficulties [Butterworth, 1999]. According to this view, humans and nonhuman animals are endowed with the ability to understand quantities and relative numerosities; mathematical abilities develop from the "innate number module." This view is supported by research suggesting that young infants demonstrate an understanding of small numerosities and simple arithmetic [Starkey et al., 1990; Wynn, 1992] and that children with MLD are particularly error prone on tasks that tap the mental representation of number. The fact that working memory deficits are not uniformly found in samples of children with MLD is also used to argue against the domain general view [Butterworth and Reigosa, 2007].

Results from our longitudinal investigation support hypotheses about the importance of certain cognitive systems such as working memory/inhibitory control in the development of math abilities, particularly when contrasted with the development of reading, but they are unable to address the question of whether domain-general skills such as working memory and inhibitory control are primarily causal in the development of mathematical ability and disability. From a developmental perspective, there are different possible pathways to math development that could explain how and when executive functions are salient. According to Zelazo et al. [2003], the knowledge of basic rule structures and problem components allows for the development of complex executive abilities. In other pathway models, inhibitory control facilitates academic skill consolidation above the influence of basic knowledge representations [Diamond et al., 2007]. The combination of these two hypotheses leads to a more circular model of math development. The early maturation of executive functions, promoted by environmental and genetic influences [Friedman et al., 2008], could enhance the early learning of basic mathematical skills (e.g., counting, small sum arithmetic). This increasingly solidified knowledge of math rule structures could facilitate the development of more advanced executive functions. In turn, these executive abilities may allow children to understand more complex mathematical concepts. Breakdowns in various stages along this circular path could lead to math deficits and perpetuate a cycle of academic underachievement. Alternatively, some integrative models suggest that a core number module interacts with domain general cognitive abilities to facilitate mathematical skill development [Spelke and Kinzler, 2007]; presumably, breakdowns in either the core number module or domain general cognitive abilities or their interaction could affect mathematical learning.

An alternative explanation for the overlap between executive functions and math has its roots in recent socio-historical trends. Historically, during the early 1900s in the US, mathematics was not offered to children in the early primary grades. At the turn of the 20th century, mathematics instruction for the upper elementary grades emphasized rote memorization and drills. Early elementary school mathematics textbooks consisted predominantly of written prose, with little exposure to diagrams or symbols. However, by the 1960s, a comprehensive mathematics curriculum had been established for all primary grades. Advanced cognitively-based math areas, like geometry, started to be introduced to younger children. Currently, math curricula continue to progressively emphasize visual-spatial concepts and executive thinking abilities [Blair et al., 2005]. These problems (e.g., pattern completion) required multiple operations and cognitive skills such as working memory. This increased utilization of cognitive executive abilities, early in a child's life, likely leads to enduring changes in both cognitive and academic functioning. It is therefore possible that children are becoming more dependent on processes such as working memory and inhibitory control to consolidate mathematical knowledge and to solve novel problems.

School-Aged Children with SB

Using low-achievement cut-offs (less than the 25th percentile), ~25% of children with SB have MLD with no co-occurring RD (MLD-only), whereas only 3% have RD without a co-occurring MLD; just over 20% have both MLD+RD and this combination of learning disabilities is often associated with socio-economic disadvantage [Fletcher et al., 2004]. The high rate of MLD-only is much higher than that in the general population [Shalev, 2007], which makes SB ideal for comparing the consequences and characteristics of MLD with and without co-occurring RD. Math difficulties in the absence of reading deficits are also apparent in other genetic disorders, including Turner syndrome, Fragile X syndrome, and 22q11 deletion syndrome (see other articles in this volume).

One subtyping model of MLD [Geary, 1993; 2004] proposes possible different pathways for MLDs depending on the presence/absence of RD. Children with both MLD and RD may have difficulties in phonological working memory that make it difficult to associate number combinations (e.g., 2 + 5) with their answers (7) to facilitate direct retrieval of math facts (2 + 5 = 7). The math problems of children with MLD-only have been variously hypothesized [Geary, 1993; Rourke, 1993] to be related to difficulties in the spatial representation and manipulation of quantities, similar to that of adults with acquired brain injuries with spatial dyscalculia [Hartje, 1987] or to difficulties in learning procedures including counting strategies in single digit arithmetic and regrouping in multidigit arithmetic. The fact that a large proportion of children with SB have MLD and co-occurring deficits in visual-spatial processing make this disorder of some interest to such subtyping models of MLD. Studies on both single and multidigit arithmetic in children with SB are presented below.

Single Digit Arithmetic

The most well studied aspect of math in children with and without MLD, including children with SB, is arithmetic. Children with SB who have MLD whether they have MLD-only or MLD+RD are slower and less accurate on single digit arithmetic problems (e.g., 4 + 3) even when they are directly retrieving the answer from memory (e.g., "I just know that 4 + 3 = 7") and they use less developmentally-mature counting strategies such as the min strategy (e.g., 4 + 5 = 5, 6, 7, 8, 9) on more problems compared to other children their age [Barnes et al., 2006]. These findings converge with those from other studies that identify difficulty in math fact retrieval as a core deficit in children with MLD [Jordan et al., 2003] and furthermore, suggest that difficulties in math fact retrieval occur in children with MLD regardless of the presence/absence of frank neurological insult or reading status. Children with SB and no learning disability had arithmetic abilities that were average, but significantly lower than those of their typically developing peers, while the pattern for word reading was just the opposite. Interestingly, these children with SB and no learning disabilities were slower in single digit arithmetic and used less mature strategies in solving these problems than their typically developing peers. Our data suggest that efficiency of math fact retrieval may be a marker of computational skill across the general population, and not simply a marker of MLD [see Barnes et al., 2006]. That is, math fact retrieval skills may be normally distributed in the population much like phonological awareness is for reading. However, unlike phonological awareness and RD, we make no claim about causal connections between math fact retrieval and MLD.

Although children with MLD and SB, regardless of reading status, were less accurate and slower on single digit arithmetic problems than were children with SB and no learning disability and typically developing age peers, there were a few differences between the groups related to reading status: the MLD-only group used more direct retrieval and solved small sum problems (sums less than 10) more quickly than the MLD+RD group. These findings are consistent with the hypothesis that children with MLD+RD show less direct retrieval of math facts because deficits in phonological working memory may disrupt the ability to learn the associations between addends and answers [Geary, 1993, 2004].

Multidigit Arithmetic

Those children with SB and MLD-only who also have deficits in visual-spatial skills related to their brain dysmorphology could be considered an ideal test of the hypothesis that there is a neurodevelopmental variant of the type of visual-spatial dyscalculia that is sometimes seen in adults with acquired brain injuries. One of the ways in which visual-spatial dyscalculia has been studied in adults [Hartje, 1987] and anecdotally reported in some children with MLD-only [Rourke, 1993] is to see whether these individuals make visual-spatial errors when solving written multidigit arithmetic problems. These include errors related to misreading and miswriting of numbers, the crowding of written work, and the like. However, on multidigit subtraction tasks, children with SB and MLD-only do not commit more visual-spatial or visual monitoring errors; that is, they were not more prone than any other group to misread or miswrite numbers, to crowd their written work, or to make errors due to misalignment of numbers in columns [Ayr et al., 2005; Barnes et al., 2006]. Nor do visual-spatial errors characterize the performance of children with SB on other operations such as multidigit multiplication and long division [Barnes et al., 2002]. Their errors in multidigit arithmetic are mainly procedural in nature reflecting less well developed conceptual and procedural arithmetic knowledge (e.g., understanding of base 10 and difficulties borrowing). In all, the findings provide no evidence for the idea that MLD without RD represents a subtype of MLD that has it origins in impairments in the spatial representation and manipulation of quantity or number.

Correlational studies tell a similar story, namely, that visual-spatial abilities are not significantly related to multidigit arithmetic performance in children with SB on either standardized or experimental tests of either single or multidigit arithmetic [Ayr et al., 2005; Barnes et al., 2006]. In contrast, like counting and counting knowledge in young preschoolers, multidigit arithmetic in school-age children with SB is related to fine motor skills [Barnes et al., 2005]. These findings are consistent with neuropsychological studies of typically developing children showing that early fine motor skills predict later achievement in mathematics [Fayol et al. 1998]. Although such findings are interesting, causal connections between the development of finger skills and math skills have not been established, nor is there solid evidence for the hypothesis that fine motor and math skills come to share common representational systems and neural substrates in parietal lobe through their developmental links [Butterworth, 1999]. Nonetheless, these findings point to ways in which theories from across the broad spectrum of psychology—in this case, neuropsychology—may be relevant to understanding MLD. That the pathophysiology of SB includes thinning of parietal lobes points to potential avenues for brain-behavior research on mathematical development in this disorder.

Although children with SB have difficulties with arithmetic, within the broad domain of mathematics, their arithmetic abilities and basic number knowledge are better developed than other aspects of mathematics such as geometry, estimation, and word problem solving. These aspects of mathematics draw to a greater extent on the neurocognitive weaknesses of individuals with SB including difficulties in manipulating visual-spatial representations [Barnes et al., 2002], and performance in these domains of mathematics is, in fact, related to visual-spatial abilities in this population as it is in typically developing children [Barnes et al., 2002].

Adults with SB

Math concepts and numerosity understanding are similarly compromised in many adults with SB [Hommet et al., 1999]. Research by Dennis and Barnes [2002] found that young adults with SB had difficulties with computation accuracy and speed, problem solving and functional numeracy involving the ability to apply numerical information to everyday situations such as making price comparisons, dealing with the value of coins, banking and budgeting, and time concepts. Furthermore, data on a subgroup of adults who had been tested as children showed that the difficulties in math are persistent across time; those individuals who scored low on an oral math problem solving task (the Arithmetic subtest of the Wechsler Intelligence Scale for Children) had considerable difficulty in similar types of math problem solving and functional numeracy as adults. The severity of these limitations was affected by working memory for numbers and lifetime number of shunt-revisions. Functional numeracy impairments limited these adults' ability to successfully complete everyday tasks related to grocery shopping, telling time, cooking, and banking. Moreover, to a greater extent than functional literacy, which was intact in these adults [Barnes et al., 2004a], functional numeracy was related to self-reported levels of social and personal autonomy. These results suggest that math deficits in SB are not the result of a developmental lag that independently resolves. Rather, these deficits are pervasive, persisting into adulthood and adversely impact quality of life. Considering the importance of mathematical competence for quality of life, adaptive functioning, and employment success [Hetherington et al., 2005], it is clear than early and persistent math impairments can have significant long-term consequences for individuals with SB.

IMPLICATIONS FOR INTERVENTION AND AVENUES FOR FUTURE RESEARCH

One of the general "rules" of research on interventions for children with academic disabilities is that gains are specific to what is taught; in other words, math and reading will not improve unless explicit teaching of math and reading content are a large part of the intervention [Fletcher et al., 2007]. Effective programs that explicitly instruct mathematical content have been reported for both preschool and school-age children [reviewed in Fletcher et al., 2007; Griffin, 2007], and there is every reason to think that such programs would also be effective for children with neurodevelopmental disorders that particularly affect the development of mathematical skills. Given the high rates of MLD in SB and some of the other neurodevelopmental disorders presented in this volume, a focus on early intervention and prevention may hold some promise; early intervention seems particularly relevant in light of the findings on persistent deficits in math that interfere with functional outcomes in adults with SB. However, one of the questions that could be posed by domain general theories of mathematical development and disability and by some of the findings reviewed above, is whether there is any role for training of executive functions and attention either alone or in combination with explicit mathematical content for preventing mathematical difficulties or for improving mathematical processing in children at risk for MLD.

Some studies suggest that regulatory abilities and executive skills such as inhibitory control, working memory, and behavioral aspects of attention may be systematically improved through training in preschoolers [Dowsett and Livesey, 2000; Diamond et al., 2007], and in school-age children with attention deficit/hyperactivity disorder [Klingberg et al., 2005]. Although correlations between performance on some executive function tasks and math were shown in the Diamond et al., study, the study design did not permit one to causally link the intervention with academic achievement. These findings suggest that specific training programs can result in generalized improvements in cognitive domains such as executive skills and attention, which are related to academic achievement, particularly in math. However, it is unknown whether such programs: (1) result in training effects that persist over time; (2) when used alone could lead to improved math outcomes, and if so, at what ages and for what types of children; (3) when used in combination with explicit mathematical content could lead to improved math outcomes over and above what is reported for those math programs alone. Conversely, one might ask whether effective preschool and school-age math interventions contain components that, in effect, "train" executive functions and attention in addition to providing mathematical content. For example, several successful math programs for children with MLD incorporate motivators to help students regulate their attention and behavior [reviewed in Barnes et al., in press].

As mentioned previously, SB is associated with brain dysmorphologies that may be particularly implicated in mathematical processing. Thinning often occurs in the posterior regions of the brain, namely the parietal and occipital cortices, of individuals with SB [Fletcher et al., 1996]. The parietal cortex, in particular, is considered to be important for several aspects of mathematical processing. For example, circuits in parietal lobe are implicated in mathematical function and dysfunction—a bilateral intraparietal system for core quantitative processing, a region of the left angular gyrus for verbal processing of numbers, and a posterior superior parietal system for mathematical processing such as estimation that may require spatial attention [Dehaene et al., 1999; 2003]. It is unknown whether similar relations between damage to parietal lobe and math hold for individuals with a neurodevelopmental disorder like SB in which the neural substrate is damaged very early in life before the development of mathematical abilities. As mentioned earlier, dysmorphology in structures such as the midbrain or parietal lobe might also affect the acquisition of mathematical skills through their effects on the development of attention and visual exploration, fine motor skills, reaching and pointing and the like. How some of these core motor and cognitive deficits in SB that are related to the primary dysmorphologies of the disorder and that are present from birth affect the development of mathematical abilities from infancy and across the lifespan would be of interest.

Functional imaging studies are currently underway in individuals with SB that should help identify particular neural networks that are implicated in different aspects of math functioning such as calculation and estimation in this neurodevelopmental disorder. Comparisons with other neurodevelopmental disorders at high risk for MLD and in children with MLD without neurodevelopmental disorder would provide knowledge about whether the neural signature of MLD varies across populations even when the behavioral phenotype looks remarkably similar. Such studies would be relevant to mathematical cognition, but also to broader questions of plasticity and neural reorganization. Finally, SB represents a potentially interesting population in which to study the behavioral and neural consequences of early intervention for mathematical difficulties. ■

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