

*Special Article***Developmental Dyscalculia**

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ABSTRACT

Developmental dyscalculia is a specific learning disability affecting the normal acquisition of arithmetic skills. Genetic, neurobiologic, and epidemiologic evidence indicates that dyscalculia, like other learning disabilities, is a brain-based disorder. However, poor teaching and environmental deprivation have also been implicated in its etiology. Because the neural network of both hemispheres comprises the substrate of normal arithmetic skills, dyscalculia can result from dysfunction of either hemisphere, although the left parietotemporal area is of particular significance. The prevalence of developmental dyscalculia is 5 to 6% in the school-aged population and is as common in girls as in boys. Dyscalculia can occur as a consequence of prematurity and low birthweight and is frequently encountered in a variety of neurologic disorders, such as attention-deficit hyperactivity disorder (ADHD), developmental language disorder, epilepsy, and fragile X syndrome. Developmental dyscalculia has proven to be a persisting learning disability, at least for the short term, in about half of affected preteen pupils. Educational interventions for dyscalculia range from rote learning of arithmetic facts to developing strategies for solving arithmetic exercises. The long-term prognosis of dyscalculia and the role of remediation in its outcome are yet to be determined. (*J Child Neurol* 2004;19:765–771).

Received December 28, 2003. Accepted for publication December 28, 2003.
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Parts of this chapter appeared in or are similar to other reviews in Shalev RS, Gross-Tsur V: Developmental dyscalculia. *Pediatr Neurol* 2001;24:337–342 (reprinted with permission from Elsevier); Shalev RS: Developmental dyscalculia, in Rapin I, Segalowitz S (eds): *Pediatric Neuropsychology*, vol 8 (part II), *Handbook of Neuropsychology*. Amsterdam, Elsevier Press, 2003, 717–729 (reprinted with permission from Elsevier); Shalev RS: Developmental dyscalculia. *Continuum Learn Disabil* 2002;8:60–73 (reprinted with permission from Lippincott Williams & Wilkins).

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Arithmetic is of prime importance in everyday life, enabling us to comprehend number concepts and perform calculations. Budgeting our time and monetary resources, reading calendars, locating an address, and even following a recipe are examples of our dependence on elementary arithmetic skills. Yet the study of the various aspects of the normal and abnormal development of arithmetic has not received the same attention as have other learning disabilities.¹ In the case of dyslexia, its neurobiology has been convincingly demonstrated, and effective remedial options have been introduced and implemented.² But for dyscalculia, these issues have been relatively neglected, and only recently have they become the subject of scientific interest. In this review, I describe the clinical, biologic, and genetic aspects of dyscalculia and the role of the physician and educator in its diagnosis and management.

DEVELOPMENT OF ARITHMETIC SKILLS

Developmental dyscalculia is defined as a specific learning disability affecting the normal acquisition of arithmetic skills in spite of normal intelligence, emotional stability, scholastic opportunity, and motivation.³ Unlike reading, which needs to be taught, children have a biologically based propensity to acquire arithmetic skills (eg, counting, adding, and comparing and understanding quantities) without formal schooling.⁴ Interestingly, the conceptual basis for numeric abilities is not exclusive to *Homo sapiens* and has been demonstrated in monkeys as well.⁵ In humans, expression of the potential for arithmetic is manifested as early as infancy by the ability of babies to discriminate between small numbers and engage in numeric computations.⁶ Although Piaget contended that young children count largely by rote and do not possess a conceptual understanding of number-related activities,⁷ preschool toddlers understand simple mathematical relations sufficient to calculate addition and subtraction for numbers up to 3.⁸ By age 3 to 4 years, they can count up to four items and, about a year later, count up to 15 and comprehend the concept that numbers represent. Eight-year-old children can write three-digit numbers, recognize arithmetic symbols, and undertake elementary exercises in addition and subtraction. Further proficiency with multiplication and division is acquired between 9 and 12 years of age.^{9–11}

CLINICAL CHARACTERISTICS

The manifestations of developmental dyscalculia are age and grade related. First-graders present with problems in retrieval of basic arithmetic facts and in computing arithmetic exercises, phenomena that presumably reflect immature counting skills.¹² Older children (9–10 years) have severe difficulties in learning arithmetic tables and comprehending algorithms of addition, subtraction, multiplication, and division. However, over the years, they have acquired basic number concepts and can write, read, or correlate the number word to its numeral. For children with dyscalculia, the solution of number problems such as $13 - 9$ or 7×6 is not self-evident. Another source of errors is their incomplete procedural knowledge of algorithms, as seen by misuse of the arithmetic sign, forgetting to carry over, misplacing digits, or undertaking the exercise in the wrong direction.^{13,14} In a case study, a dissociation between knowledge of number facts and arithmetic procedures was documented, illustrating the paradox between the ability to master number facts and the inability to solve a complex arithmetic exercise.¹⁵

PREVALENCE AND EPIDEMIOLOGY

Population studies in countries as diverse as the United States, Germany, India, and Israel demonstrate that the prevalence of developmental dyscalculia ranges from 3 to 6.5%.^{13,16–19} This evidence-based information is yet to be incorporated into the *Diagnostic and Statistical Manual of Mental Disorders-IV*, which states that developmental dyscalculia is rare, with a prevalence of only 1%.³ The number of girls with dyscalculia is equivalent to that of boys, an unexpected finding considering that learning disabilities are generally more prevalent in boys. In general, dyscalculia appears as an isolated and specific learning disability; however, about one quarter have comor-

bidity for attention-deficit hyperactivity disorder (ADHD) and dyslexia.^{13,19} Compared with children with developmental dyscalculia alone or those with dyscalculia and ADHD, children with dyscalculia in combination with dyslexia are more profoundly impaired.¹⁴

PROGNOSIS

There is little information about the short-term follow-up of developmental dyscalculia, and virtually no data are available concerning its long-term outcome. It has to be emphasized, however, that because learning occurs over a variable spectrum of time, not every delay in acquiring a cognitive skill indicates a learning disability. Indeed, arithmetic difficulties in the first grades of elementary school can reflect such a developmental variability. Not surprisingly, children with dyscalculia in first and second grades show considerable improvement in their ability to master counting procedures over a short period of time.¹² However, when fifth grade boys and girls with developmental dyscalculia were prospectively followed and re-examined 3 years later, their prognosis was not favorable: 95% achieved scores falling in the lowest quartile for their school class and 47% in the lowest 5th percentile. Thus, the severe form of dyscalculia, the criteria of which are consistent with the diagnosis of a specific learning disability in arithmetic, persists in as many as 50% of teenage children.²⁰ Factors associated with persistence of dyscalculia were the severity of the arithmetic disorder at the time of initial diagnosis and the presence of arithmetic problems in the siblings. Factors not associated with persistence of developmental dyscalculia were socioeconomic status, gender, co-occurrence of another learning disability, and the extent or types of educational interventions received.²⁰

ETIOLOGY AND NEUROBIOLOGY

The etiology of developmental dyscalculia is most probably multifactorial, including genetic predisposition, environmental deprivation, poor teaching, classroom diversity, untested curricula, mathematical anxiety, and neurologic deficits. The role of genetics has been demonstrated convincingly in a twin study paradigm, which showed that monozygotic and dizygotic cotwins of dyscalculic probands were 12 and 8 times, respectively, more likely to also have dyscalculia than the normal population.²¹ Moreover, in a study of 33 families, approximately half of all siblings of probands with dyscalculia were dyscalculic, with an estimated risk that was 5 to 10 times greater than expected.²² These studies highlight the role of heredity in the etiopathogenesis of dyscalculia, postulated by Kosc some 25 years ago.²³

It is important to realize that children have an inherent ability to learn some arithmetic on their own, but, for the most part, this skill is acquired through schooling. Therefore, assessing the adequacy of instruction is relevant in the diagnosis of children whose arithmetic achievement is below that expected for age and grade. Also, many arithmetic curricula are empiric, often chosen by educators without sufficient evidence as to their efficacy.²⁴ Furthermore, teachers might move on to new material before ascertaining competency in concepts previously taught.²⁵ Compounding drawbacks are overcrowded classrooms, the diversity of the student body, and a trend toward mainstreaming.²⁴

Mathematical anxiety can be both the cause of and symptomatic to dyscalculia. Children with mathematical anxiety tend to sacrifice accuracy for speed, performing poorly even on the most basic arithmetic exercises.²⁶ Fortunately, psychologic intervention for mathematical anxiety can improve competence in arithmetic.²⁷ Whether pharmacotherapy in this condition is useful awaits documentation.

The application of sophisticated electrophysiologic studies and neuroimaging techniques has yielded valuable information on the neuroanatomic basis of arithmetic. Using event-related potentials, Kiefer and Dehaene found that simple multiplication exercises are processed by the left parietal cortex, whereas complex exercises are executed within both centroparietal areas, albeit more so on the left.²⁸ Neuroimaging by functional magnetic resonance imaging (MRI) in normal individuals engaged in arithmetic shows bilateral activation of prefrontal and inferior parietal cortices.²⁹ When the arithmetic task is an exact, language-dependent calculation (eg, "Seven times five is..."), a large area in the left inferior frontal lobe is activated. On the other hand, tasks of number approximation (eg, "Which is larger, 5 or 7?") activate both parietal lobes.³⁰ Elegantly executed experiments indicate that there are two neighboring regions within the left intraparietal sulcus that are activated during subtraction. Its middle portion is activated exclusively during subtraction, whereas the more posterior region is activated by both phonemic and subtraction tasks. This duality is consistent with the theory that number processing relies on verbal and nonverbal representation of numbers.³¹

Functional neuroimaging in dyscalculia has only recently begun to emerge. In one young adult with dyscalculia and no known structural abnormalities, magnetic resonance spectroscopy demonstrated a wedge-shaped defect in the left parietotemporal area.³² In another patient with dyscalculia secondary to a right temporal lobe hemorrhage endured at infancy, functional MRI activation was localized predominantly to the frontal and parietal regions of the intact left hemisphere.³³ We performed functional MRI in four young adults with developmental dyscalculia who, when shown an arithmetic exercise, were required to estimate whether the suggested response was close to or distant from the correct answer. We found that brain areas activated by the dyscalculic patients were the right intraparietal sulcus and the left middle frontal gyrus. Controls, on the other hand, activated the intraparietal sulcus bilaterally with minimal activation of the left frontal gyrus.³⁴ Functional MRI in two sets of twins, of whom only one cotwin had dyscalculia, demonstrated that the dyscalculic twins activated large areas of the brain, including the frontal, precentral, and dorsal parietal areas. However, their twins, like normal children, activated predominantly the left frontal and temporoparietal areas.³⁵ In an elaborate functional MRI study in 14 women with Turner's syndrome and dyscalculia, the expected activation of parietal areas during arithmetic calculations failed to occur. Morphologic analyses of the parietal areas showed anatomic disorganization of the right intraparietal sulcus, an area already identified as critical for arithmetic functioning.³⁶ Although preliminary, these data suggest that individuals with dyscalculia can activate alternative brain areas. It is intriguing to theorize that children and young adults with dyscalculia are unable to activate regions that are normally operant during arithmetic exercises because these areas are

dysfunctional or dysmorphic, as in the case of Turner's syndrome. In their effort to compensate for the disability, individuals with dyscalculia recruit other brain regions, employing substitute but, unfortunately, inefficient cognitive strategies.

THEORETIC MODELS

In 1925, Henschen coined the term "acalculia" to denote the loss of previously acquired arithmetic skills. He postulated the existence of a distinct and autonomous cortical network for arithmetic and perceived acalculia as a manifestation of a specific neurocognitive deficit.³⁷ Shortly thereafter, Berger demonstrated that acalculia could also occur within the context of a general cognitive decline.³⁸ Hecaen et al attributed dyscalculia to three neurobehavioral impairments: agraphia and alexia for numbers, spatial dyscalculia, and anarithmetia.³⁹ Benson and Denckla subsequently described a fourth potential neurobehavioral mechanism, termed "number paraphasia."⁴⁰

These and other neuropsychologic deficits comprised the conceptual basis for the underpinnings of developmental dyscalculia. Rourke, for example, proposed that dyscalculia is secondary either to visuospatial or to verbal and auditory-perceptual dysfunction, both of which could culminate in multiple cognitive manifestations, including dyscalculia.⁴¹ Poor working memory, including spatial working memory, has also been proposed as underlying developmental dyscalculia.^{42,43} In fact, Geary et al attributed the failure of children to develop long-term memory representations of number facts to working memory deficits.⁴⁴ Defective inhibitory mechanisms can also interact with working memory; specifically, if working memory is improperly modulated, irrelevant information or inappropriate arithmetic strategies are not inhibited in favor of more suitable choices.⁴⁵ Speed of processing deficits has also been implicated, leading to a lack of automaticity of basic arithmetic facts.⁴⁶

Temple and Sherwood challenged the notion that dysfunction of an underlying neuropsychologic process is responsible for arithmetic problems.⁴⁷ They demonstrated that retrieval of arithmetic facts is a specialized function, not linked to short-term memory or impairments in other cognitive domains. Their data are consistent with other experimental evidence that arithmetic skills are inherent, present from infancy, enabling babies to identify and distinguish small quantities of numbers and objects.^{6,48,49} These observations imply that arithmetic processing is dependent on a specialized cognitive mechanism rather than a product of "general" neuropsychologic processes.^{6,47}

Another approach to the complexities of arithmetic is the neurocognitive models that attempt to explain both normal arithmetic processing and dyscalculia.^{50,51} Accordingly, the model developed by McCloskey et al divides arithmetic skills into three main groups: comprehension of number concepts, production of numbers, and calculation. It provides a theoretical basis to explain isolated deficits in a specific domain of arithmetic while other facets of arithmetic remain intact.⁵⁰ Such a dissociation was reported by Temple, who described a boy with developmental dyscalculia whose deficit was restricted to lexical number processing.⁵² Although this boy had no difficulty in reading simple or unfamiliar words, he was unable to attribute the precise word

value to a digit. Thus, when called on to read the number "9172," he read it as "six thousand, six hundred and seventy-two." The magnitude of each digit was correctly identified, although individual numerals were misread. Additional support for this model is derived from children with dyscalculia and isolated deficits in learning arithmetic tables or inability to execute arithmetic procedures.¹⁵

The "triple-code model" proposed by Dehaene and Cohen has three components: verbal, visual, and magnitude.⁵¹ Accordingly, relatively simple arithmetic operations are processed by the verbal system within the left hemisphere, whereas complex arithmetic procedures—which require magnitude estimation and visual representations—are bilaterally localized. This model is supported by neuroimaging from normal individuals and by case studies of patients with focal brain lesions. Thus, when normal individuals perform overlearned arithmetic exercises, which are presumably language dependent, functional MRI shows left frontal lobe activation. Solving complex calculations, which require combined visual and numeric magnitude representations, activates both parietal lobes.^{29,30} In a clinical case study, Grafman et al described a patient with a left hemisphere lesion who could no longer remember rote, overlearned arithmetic facts but demonstrated intact knowledge of number magnitude.⁵³ Based on these data, Dehaene et al proposed that the arithmetic neural network is composed of two distinct neural circuits, linguistic and visual-spatial, and concluded that both are necessary for arithmetic processing.³⁰ Depending on the extent and severity of the deficit to adjacent neural networks, developmental dyscalculia can appear either as an isolated learning disability or in conjunction with other learning and neurologic problems, such as dyslexia, dysgraphia, and epilepsy.^{13,54}

DIAGNOSIS

Although epidemiologic, neurobiologic, and genetic evidence indicates that the underpinnings of learning disabilities are brain based,^{2,55} their diagnosis is still predicated on the assessment of academic skills. In fact, conventional medical and neurologic examinations generally add only marginally in the diagnostic process. When dyscalculia is suspected, the first step is to assess the child's arithmetic skills. The diagnosis is substantiated if there is a significant discrepancy between the intellectual potential of the child and his arithmetic achievement or by a discrepancy of at least 2 years between the chronologic grade and the child's level of achievement.⁵⁶ However, there are caveats to both definitions. The latter has limited usefulness in cases in which a 2-year discrepancy is not meaningful, that is, in the very first grades of school or for those completing high school. The reservation regarding the achievement potential definition is that gifted children whose achievement is still within the normal range, albeit significantly lower than expected, will be identified as learning disabled.

Standardized arithmetic tests are the acceptable method to assess arithmetic skills in children. The arithmetic subtests of the Wide Range Achievement Test—Revised and the Young's Group Mathematics Test are timed tests that emphasize achievement.^{57,58} Recently, a battery of arithmetic tests called the Neuropsychological Test Battery for Number Processing and Calculation in Children (NUCALC) was designed to assess number concepts, number facts, and arithmetic procedures and

was validated for Switzerland and France.^{59–61} We developed and validated an arithmetic test for elementary and high school pupils. The test, based on the neurocognitive model of McCloskey et al,⁵⁰ assesses number concepts and arithmetic procedures and is useful for research purposes.^{11,20,22}

Clinicians who provide comprehensive care should be aware that dyscalculia is not only a very common learning disability but also the most frequently encountered cognitive problem in a variety of medical conditions. Children born preterm at low and very low birthweights are at high risk of developing dyscalculia.^{62,63} This is also the case for children with epilepsy⁶⁴ or genetic disorders of the X chromosome (ie, Turner's syndrome)⁶⁵ and girls with fragile X syndrome.⁶⁶ It can also occur in the context of other developmental cognitive disorders, such as developmental dysphasia, ADHD, Gerstmann's syndrome, and developmental dyslexia.^{67,68} In fact, kindergarten children with developmental language disorders frequently manifest impaired arithmetic skills, and over 20% of boys with ADHD have dyscalculia.^{69,70} They make numerous mistakes in arithmetic, possibly owing to impaired recall, careless errors, inattention to detail, and overall slowness.⁷¹ We therefore advise that before giving a child with ADHD the diagnosis of dyscalculia, the ADHD should be adequately treated and arithmetic skills subsequently re-evaluated.

TREATMENT

Children with learning disabilities are vulnerable to multiple risks, including persistence of the learning handicap, school dropout, and emotional instability; children with dyscalculia apparently face similar challenges.^{20,72} Therefore, treatment of dyscalculia should address the multiple facets of the disorder while focusing on educational interventions to improve study skills in general and strengthening number perception and arithmetic concepts in particular. Research in this domain indicates that students with learning disabilities can improve their overall study skills and benefit from specific techniques for their individual problem.^{72,73} Nonacademic skills that have been coined "classroom survival skills" are essential to all students, especially those with learning disabilities. Acquisition of survival skills, examples of which are assuming accountability for one's own behavior, arriving prepared for lessons, meeting deadlines, appropriate school behavior, following directions, and completing homework assignments, becomes indispensable with increasing age and mounting responsibilities. Intervention programs, designed to enhance these skills, have long-term beneficial effects.⁷³

A variety of educational techniques for children with arithmetic disabilities have been proposed. For those unable to decipher the syntactic elements of numbers, a training program to transcribe numbers into their corresponding digits has been developed. Thus, for the number "60,349," the student practices number syntax by learning that the "6" relates to tens of thousands, the "0" to thousands, the "3" for hundreds, and so forth.⁷⁴ The ability to internalize the concept of "number line" can be taught by repetitive additions, for example, using 10, that is, $4 + 10 = 14$, $14 + 10 = 24$, $24 + 10 = 34$, etc.⁷⁵ Rourke and Conway suggested that remediation focus on the neuropsychologic problem underlying the dyscalculia, whether it is perceptual and visual-spatial or verbal and auditory-

perceptual.⁷⁶ This approach stresses verbalization of arithmetic concepts, procedures, and operations.

The importance of interactive learning in arithmetic is exemplified by the MASTER (Mathematics Strategy Training for Educational Remediation) program developed for teaching multiplication and division. It conveys the principle of these two arithmetic operations (eg, that multiplication is repeated addition) while learning how to choose appropriate strategies (eg, 7×6 could be solved by adding 7 six times or by adding the results of 5×6 and 2×6). The efficacy of this program suggests that children with dyscalculia can learn arithmetic when provided with number concepts and problem-solving strategies.^{77,78}

Children who understand number concepts but have difficulty in computation can be overwhelmed by the procedures needed to solve an arithmetic exercise. Therefore, enhancing automatic recall for number facts by drilling can also be helpful. The theoretic basis for the importance of automaticity stems from evidence that children with poor mathematical skills have deficits in speed of processing and inhibition.⁴⁵ Thus, implementing programs that promote rote learning of arithmetic facts needs to be reconsidered.²⁵

Gersten and Chard argued that internalizing the concepts of “number sense,” that is, the intuitive knowledge of numbers and procedures needed for numeric operations, is crucial in remediation of dyscalculia.¹ Accordingly, children should be instructed in basic number concepts rather than rote memorization of number facts. They suggest using representational systems to facilitate learning, an example of which is a thermometer for the concept of number line. Another method to promote concept understanding is to encourage students to verbalize their perception of the arithmetic procedure while the teacher provides feedback. Similarly, enhancing planning skills specific for arithmetic has proved beneficial in instructing children with dyscalculia.^{78,79}

Assistive technology should also be available for the dyscalculic child. Software for mathematics that emphasizes drill has the potential to aid learning number facts. Interactive video disc programs for acquiring math concepts and problem-solving skills, as well as for providing instruction for everyday situations dependent on mathematical thinking, are now available.⁸⁰ Pocket calculators are helpful when impaired memorization of number facts impedes the ability to correctly complete an arithmetic problem.⁴ Calendars, time management programs, and digital and talking clocks should also be recommended.

When all else fails, the option of a “time-out” period should be considered with the anticipation that a developmental change will occur, enabling better comprehension of arithmetic concepts. In the absence of such maturation, as is often seen in teenagers with dyscalculia, it is imperative to ascertain that knowledge of arithmetic concepts required for daily life (eg, street address, calendars, money, and checks) has been adequately learned.

The role of the physician in the management of dyscalculia begins in the diagnostic phase by establishing whether the learning disability is a primary, symptomatic, or comorbid entity. Physicians, with their inherent authority, are often the appropriate member within the multidisciplinary team to discuss with parents the nature and ramifications of the cognitive disability affecting their child. They can guide both child and parents through the maze of expert assessments and multitude of therapeutic options while

providing explanations regarding the neurologic underpinnings of the disorder and its familial-genetic implications.

FUTURE DIRECTIONS

Research in dyscalculia is still in its emergent stage, lagging behind other learning disabilities. Although theories are plentiful, the underlying neuropsychologic process or deficit(s) of dyscalculia remains elusive. Functional neuroimaging and genotyping of individuals with dyscalculia and their families might provide clues for ultimately identifying its structural, genetic, and molecular biology. The impact of environment on the severity of dyscalculia and its chronicity and comorbidity also awaits further investigation.

CASE REPORTS

Case 1

A 14-year-old boy in grade 9 was referred to the neuropsychiatric unit because of ADHD and an inability to learn basic concepts of arithmetic. His family history was positive for an older sister with dyscalculia. The diagnosis of ADHD was already determined in the second grade, and he responded to pharmacologic therapy with psychostimulants. He succeeded in learning reading and writing but lagged behind his classmates in arithmetic such that in third grade, he was still using his fingers to add or subtract. Intensive remediation was begun, and he eventually mastered complex addition and subtraction, without adequately comprehending or automatizing the procedures of multiplication. To solve 7×5 , he would repetitiously add 7 five times but could not grasp the concepts necessary for division. His physical and neurologic examinations were normal, as were his Wechsler Verbal IQ (107) and Performance IQ (110).

This case demonstrates the occurrence of developmental dyscalculia and ADHD, a comorbid situation found in a quarter of children with dyscalculia. The presence of a sibling with dyscalculia points to the familial-genetic nature of this disorder. Although our patient had no significant problems with reading and writing, his arithmetic remained problematic despite educational interventions and effective treatment of ADHD.

Case 2

An 11-year-old girl in grade 5 was referred to the neuropsychiatric unit because of an inability to learn the multiplication tables and arithmetic procedures necessary for multiplication and division. There was no family history of learning disabilities or ADHD, and her prenatal, natal, and psychomotor development was normal. Her initial integration into the school framework was smooth, and she acquired reading and writing skills as expected. However, in grade 3, a sharp decline in her overall behavior and school functioning was noted and attributed to an anxiety reaction following a short separation from her parents. She responded erratically to minor changes in her daily schedule, avoided unfamiliar situations, demanded parental presence at bedtime, and refused to see classmates. Her teachers reported that her arithmetic skills were very poor, both in absolute terms and relative to her verbal abilities. Although she had mastered addition and subtraction, she had no idea how to proceed in multiplication or division exercises and had not mastered the multiplication table. Psychologic and educational interventions yielded minimal improvement.

The physical and neurologic examinations were normal, as were her Wechsler Verbal IQ (110) and Performance IQ (119). Although her reading skills were age appropriate, she had difficulty in reading comprehension and poor organizational skills but no overt signs of inattention. The scores on the Child Behavior Questionnaire were in the clinical range for the anxiety/depression, withdrawal, and thought disorder scales.⁸¹ The results on a continuous performance task were well within normal limits. The working diagnosis was comorbidity for developmental dyscalculia and anxiety, although it was not apparent if the dyscalculia was symptomatic to the anxiety disorder or vice versa. The consulting psychiatrist advised treatment

with fluvoxamine for the anxiety disorder; arithmetic tutoring was also started. Within 6 months, a dramatic improvement in the symptoms of anxiety occurred, and she rapidly overcame the gap in arithmetic. This case demonstrates that dyscalculia can be symptomatic to an anxiety disorder and can respond to specific therapy.

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