Developmental dyscalculia is a specific learning disability affecting the acquisition of arithmetic skills in an otherwise-normal child. Although poor teaching, environmental deprivation, and low intelligence have been implicated in the etiology of developmental dyscalculia, current data indicate that this learning disability is a brain-based disorder with a familial-genetic predisposition. The neurologic substrate of developmental dyscalculia is thought to involve both hemispheres, particularly the left parietotemporal areas. Developmental dyscalculia is a common cognitive handicap; its prevalence in the school population is about 5-6%, a frequency similar to those of developmental dyslexia and attention-deficit–hyperactivity disorder. Unlike these, however, it is as common in females as in males. Developmental dyscalculia frequently is encountered in neurologic disorders, examples of which include attention-deficit–hyperactivity disorder, developmental language disorder, epilepsy, and fragile X syndrome. The long-term prognosis of developmental dyscalculia is unknown; it appears, however, to persist, at least for the short-term, in about half of affected preteen children. The consequences of developmental dyscalculia and its impact on education, employment, and psychologic well-being of affected individuals are unknown. © 2001 by Elsevier Science Inc. All rights reserved.


Introduction

Developmental cognitive disorders in general and learning disabilities in particular have long been perceived outside the pale of classic neurology, expressing perturbations related to the mind rather than to the brain [1]. However, over the past two decades sufficient genetic, neurobiologic, and epidemiologic evidence has accumulated to indicate that learning disabilities are in fact expressions of brain dysfunction. However, the role of the physician in the diagnosis and treatment of these disorders has not been self-evident. This observation is best illustrated by the fact that conventional medical and neurologic evaluations add only limited information to patients with attention-deficit–hyperactivity disorder (ADHD), developmental dyslexia, and developmental dyscalculia. Indeed, the diagnosis of these disorders usually is predicated on information gleaned from the history, age-appropriate assessments of academic skills, and standardized behavioral questionnaires. Thus although learning disabilities currently are perceived as biologically based neurologic disorders [2,3], their diagnosis is made primarily by nonmedical methods. What then, does medicine have to offer the large number of children with learning disabilities—estimated at 4-5% of the school-age population—who may turn to their physician with these problems [4]?

In this review, the clinical characteristics of developmental dyscalculia, its etiology and neurobiology, diagnosis, and treatment will be described, and the role of the physician in this common learning disability are discussed.

Arithmetic and Developmental Dyscalculia

Children exhibit a biologically based propensity to acquire arithmetic skills [5]. Unlike reading, which needs to be taught, certain numerical skills, for example, counting, adding, comparing and understanding quantities, develop naturally without formal schooling [5]. This numerical capacity is thought to be an inherent trait, present as early as infancy, exemplified by the ability of infants to discriminate between small numbers and engage in numerical computation [6]. In cultures where schooling is the norm, preschool toddlers understand simple mathematical relations and correctly add and subtract numbers up to three [7]. By 3-4 years of age, they can count up to four items, and, about a year later, count up to 15, as well as comprehend the concept that numbers represent. Eight-
year-old children can write three-digit numbers (without reversing integers or inserting extra zeroes), recognize arithmetic symbols, and perform elementary exercises in addition and subtraction. Further proficiency in addition, subtraction, multiplication, and division is acquired between 9 and 12 years of age [8-10].

Children who present with difficulty in learning arithmetic and who fail to achieve adequate proficiency in this cognitive domain despite normal intelligence, scholastic opportunity, emotional stability, and necessary motivation have developmental dyscalculia [11]. Some have trouble learning the arithmetic tables; others never comprehend algorithms of addition, subtraction, multiplication, and division; whereas others either have problems understanding the concept of numbers or cannot write, read, or identify the correct word to the numeral.

Clinical Characteristics

The manifestations of developmental dyscalculia are age and grade related. First graders with developmental dyscalculia present with problems in the retrieval of basic arithmetic facts and in the computation of arithmetic exercises, phenomena that presumably reflect their immature counting skills [12].

The manifestations of developmental dyscalculia are different in older children (9-10 years of age). By this age, they have finally mastered counting skills and are also able to match written arabic numerals to quantities; to understand concepts such as more, less, and equivalence; to recognize the numerical value of numerals; to determine which of a pair of numerals is greater or smaller; to correctly arrange numbers in serial order both from lowest to highest and vice versa; and to copy and write numbers to dictation [13]. Most children also become proficient in handling money, understanding calendrical knowledge, and using checks. At this age group, developmental dyscalculia is characterized by deficits in the retrieval of overlearned information. The solutions to number facts such as $13 - 9$ or $7 \times 6$ are not readily available to these children who, in an attempt to bypass their difficulty in solving arithmetic problems, will use inefficient strategies. Consequently they commit errors resulting from incomplete procedural knowledge necessary for complex exercises in addition, subtraction, multiplication, or division. These errors include inattentiveness to the arithmetic sign, use of the wrong sign, forgetting to “carry over,” misplacement of digits, or undertaking the arithmetic exercise in the wrong direction [13,14]. There are reports of children in whom the developmental arithmetic problem was characterized by a dissociation between arithmetic fact abilities and procedural skills, an example of which is the child who mastered number facts but didn’t know which steps were needed to solve the arithmetic exercise [15].

Prevalence and Epidemiology

According to the DSM-IV, developmental dyscalculia is a rare learning disability, with a prevalence of 1% in the school-age population [11]. However, a more realistic estimate is 5%, which is similar to that of developmental dyslexia and ADHD. Population studies for the United States, Europe, and Israel demonstrate that the prevalence of developmental dyscalculia in these countries is similar, ranging from 3-6.5% [13,16-18]. The 6.5% estimate was obtained after a two-stage screening process. In the first stage, all 10 to 11 year olds ($n = 3029$) underwent a city-wide arithmetic achievement test assessing counting skills, number facts, complex arithmetic exercises, and word problems. Those children scoring in the lowest 20% on this arithmetic test entered the second stage, whereupon they were tested on an individually administered, standardized arithmetic battery. A child was classified as dyscalculic if his or her full-scale intelligence quotient was equal to or less than 80 and when the score achieved on the second arithmetic test was equal to or less than the mean score for normal children two grades younger [13]. The final cohort, composed of 140 children with dyscalculia, was characterized by a slightly higher prevalence of females than males (1.1:1), an unexpected finding considering that learning disabilities in general are at least threefold more prevalent in males than in females [11]. Furthermore, in the majority of the cohort, developmental dyscalculia was an isolated learning disability. However, 17% of the children also had dyslexia, and 26% had ADHD. Interestingly, children who were comorbid for dyslexia were impaired more profoundly on arithmetic skills and neuropsychologic tests than children with developmental dyscalculia alone or developmental dyscalculia and ADHD [14].

Prognosis

There is little information about the short-term follow-up of developmental dyscalculia, and virtually no data are available concerning the long-term outcome of this common learning disability. However, some lessons can be extrapolated from the experience with dyslexia. Longitudinal studies of dyslexia indicate that it is a long-lasting problem and not merely an expression of a developmental lag [19,20]. Developmental dyscalculia may prove to have a similar natural history. Between the first and second grade, children with developmental dyscalculia demonstrate considerable improvement in their ability to master counting procedures [12]. However, when 10- to 11-year-old males and females with developmental dyscalculia were prospectively monitored and re-examined 3 years later, their arithmetic achievement was still poor, and their scores fell in the lowest quartile of their school class. Moreover, 47% of the children who were given the diagnosis of developmental dyscalculia in fifth grade continued to exhibit dyscalculic phenomena while in
Etiology and Neurobiology

To date no consensus as to the etiology of developmental dyscalculia has been reached. Possible contributing factors as diverse as genetic predisposition, neurologic abnormalities, and environmental deprivation all have their proponents. The role of genetics in children with dyscalculia has been investigated using a twin study paradigm. Alarcon et al. [22] found that 58% of monozygotic co-twins and 39% of dizygotic co-twins of dyscalculia probands had developmental dyscalculia and the concordance rates were 0.73 and 0.56, respectively. Using a family study paradigm, Shalev et al. [23] demonstrated that approximately half of all siblings of children with developmental dyscalculia are also dyscalculic, the risk being 5-10 times greater than for the general population. These results highlight the role of heredity in the etiopathogenesis of dyscalculia first postulated by Kose [24] some 25 years earlier.

The neuroanatomic basis of arithmetic is yet to be unraveled, although application of electrophysiology and neuroimaging techniques are yielding encouraging information. Using event-related potentials, Kiefer and Dehaene [25] found that simple multiplication is processed by the left parietal cortex, whereas complex exercises are executed within both centroparietal areas, albeit slightly greater on the left. Neuroimaging studies support these electrophysiologic findings: in normal individuals engaged in arithmetic, functional magnetic resonance imaging (fMRI) reveals bilateral activation of prefrontal and inferior parietal cortices [26]. When the arithmetic task is an exact, language-dependent calculation (e.g., “seven times five is . . .”), a large area in the left inferior frontal lobe is activated. On the other hand, tasks of number approximation (e.g., “Which is larger, five or seven?”) activate both parietal lobes [27]. Thus far, functional neuroimaging has been used to study dyscalculia in only two patients. The first involved an adolescent with dyscalculia secondary to a right temporal lobe hemorrhage endured at infancy. His fMRI activation pattern was predominantly unilateral, localized to the frontal and parietal regions of the intact left hemisphere [28]. In the second patient, a young adult with developmental dyscalculia with no known structural abnormalities, magnetic resonance spectroscopy demonstrated a wedge-shaped defect in the left parietotemporal area [29]. Taken together, the data derived from normal individuals during arithmetic processing and the pathologic findings in patients with dyscalculia suggest that this cognitive skill is predicated on a neural network involving anterior and posterior areas of both hemispheres of the brain.

However, not all researchers agree that developmental dyscalculia is a genetic, biologically based brain disorder. Other etiologies implicated in its genesis are environmental deprivation [30], poor teaching, low intelligence [31], and mathematical anxiety [32].

It is important to realize that although children can learn some arithmetic on their own, for the most part this skill is taught in school. Within this formal setting, inadequate teaching methods may be one of the reasons why children have trouble learning arithmetic. Many arithmetic curricula are empiric, often chosen by educators without sufficient evidence as to the effectiveness of a particular text [31]. Other drawbacks are overcrowded classrooms, the diversity of the student body, and a trend toward mainstreaming [5]. The latter situation generally means that children with difficulties will be receiving much of their instruction in mainstream classrooms rather than in resource rooms geared to better address their individual capabilities and specific disabilities [31].

Mathematic anxiety may masquerade as or exacerbate dyscalculia because individuals with this problem tend to sacrifice accuracy for speed, and their performance is poor even on the most basic arithmetic exercises [32]. Psychologic intervention for mathematical anxiety significantly improves competence in arithmetic [33].

Theoretical Models

In 1925, Henschen [34] coined the term acalculia, postulating the existence of a distinct and autonomous cortical network for the execution of arithmetic functions. Thus acalculia was perceived as a manifestation of a specific neurocognitive deficit. Shortly thereafter Berger [35] demonstrated that acalculia, although indeed a specific cognitive deficit, could also be part of a larger clinical spectrum, including disturbances in memory and language. He denoted the former primary acalculia and the latter secondary acalculia [35]. Hecaen et al. [36] correlated problems in performing calculations to three neurobehavioral impairments: agraphia or alexia for numbers, spatial dyscalculia, and anarithmetia; whereas Benson and Denckla [37] later described yet another subtype of dyscalculia secondary to number paraphasia.

Current neurobehavioral deficits hypothesized to be operative in developmental dyscalculia are predicated, for the most part, on the above concepts. Rourke [38], for example, proposed that dyscalculia is secondary either to visuospatial or to verbal and auditory-perceptual dysfunction, both of which can result in multiple cognitive manifestations, only one of which is dyscalculia. Poor working memory has also been proposed as a neuropsychologic mechanism underlying developmental dyscalculia [39]. It is not clear, however, how these hypotheses can be reconciled with existing experimental evidence indicat-
ing that arithmetic skills are already present in infants, enabling them to identify quantities of up to four, reason about numbers and objects, and distinguish between larger and smaller quantities [6,40,41]. These latter observations imply that arithmetic processing is basically an inherent skill, dependent on a specialized cognitive mechanism, rather than a product of “general” neuropsychologic processes [6].

Two neurocognitive models have been proposed to explain both normal arithmetic processing and dyscalculia [42,43]. The model developed by McCloskey et al. [42] divides arithmetic skills into the following three main groups: (1) comprehension of number concepts; (2) production of numbers; and (3) calculation. It provides a theoretical basis to explain isolated deficits in a specific domain of arithmetic, whereas other facets of arithmetic function remain intact. An example of such a dissociation was reported by Temple [44], who described a male with developmental dyscalculia who had an isolated deficit in lexical number processing. Although this male had no difficulty in reading simple or unfamiliar words, he was unable to attribute the precise word value to a digit. Thus, when the child was 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 descriptions of isolated deficits in learning arithmetic tables and from an inability to execute arithmetic procedures described in children with developmental dyscalculia [15].

The “triple-code model” proposed by Dehaene and Cohen [43] is both neuropsychologically and anatomically based. The three elements are verbal, visual, and magnitude representation. According to this model, relatively simple arithmetic operations are processed by the verbal system within the left hemisphere, whereas more complex arithmetic procedures—which require magnitude estimation and visual representations—are bilaterally localized. This model is supported by experimental data from normal individuals performing arithmetic, as well as by case reports of patients with focal brain lesions [43]. For example, Grafman et al. [45] described a patient with a left hemisphere lesion who could no longer remember rote, overlearned arithmetic facts but demonstrated intact knowledge of number magnitude. Data from normal individuals are based on fMRI and electrophysiologic studies [27]. Thus when normal individuals perform overlearned arithmetic exercises, which are presumably language-dependent, the activation pattern on fMRI is localized to the left frontal lobe. When the calculations are more complex, requiring combined visual and numerical magnitude representations, both parietal lobes are activated [8,26]. Based on these data, Dehaene et al. [27] proposed that the arithmetic neural network is composed of two distinct neural circuits, linguistic and visual-spatial, both of which are necessary for arithmetic processing. From a clinical perspective, one could hypothesize that the profile of arithmetic dysfunction is a function of the localization and/or severity of the damage to the arithmetic neural network. Depending on the extent and severity of the deficit to neural networks, developmental dyscalculia can appear either as an isolated learning disability or together with other learning and neurologic problems, such as dyslexia or dysgraphia [13,46].

**Diagnosis and Treatment**

The diagnosis of developmental dyscalculia is based on assessment of the child’s arithmetic skills. This assessment is determined either by the discrepancy between the intellectual potential of the child and his or her arithmetic achievement or by a discrepancy of at least 2 years between the chronologic grade and the level of achievement [47,48]. The latter definition has limited usefulness for both younger children and older individuals, in whom a 2-year discrepancy is not meaningful. The achievement-potential definition, on the other hand, will identify children of low intelligence whose performance in arithmetic is significantly lower than expected for the child’s aptitude. It will also identify gifted children whose achievement is still within the normal range, albeit significantly lower than expected. However, specific learning disabilities are not the only reason why a child, gifted or otherwise, may not perform according to aptitude. It is ultimately the clinician who will determine which child in this category has a bona fide learning disability meriting interventions [49]. It is therefore important for physicians caring for children with learning disabilities to know how the diagnosis was reached. Depending on the diagnostic method used, a child may or may not be eligible for educational benefits offered by school or government bodies [50].

Standardized arithmetic tests are the acceptable method to assess arithmetic skills in children. The arithmetic subtests of the WRAT-R [51] and the Young’s Group Mathematics Test [52] are timed tests that emphasize arithmetic achievement. Recently a battery of arithmetic tests called Neuropsychological Test Battery for Number Processing and Calculation in Children was developed [53]. The test battery, validated for grades 2-4 in Switzerland and France, was designed to assess number concepts, number facts, and arithmetic procedures [54,55]. We developed and validated an arithmetic test based on the neurocognitive model of McCloskey et al. [42]; the test assesses number concepts and arithmetic procedures and has proven useful for research purposes [10].

Different educational techniques for children with arithmetic disabilities have been proposed. Rourke and Conway [56] suggested that remedial education should focus on interventions appropriate for the underlying neuropsychologic problem of the child, for example, perceptual and visuospatial or verbal and auditory-perceptual. For children in whom memorization of number facts impedes their ability to complete the solution of an arithmetic problem,
the use of pocket calculator may be helpful [5]. Other recommendations are a “time-out” period until or if a developmental change occurs in the young child thereby allowing for better comprehension of arithmetic concepts. The management of dyscalculia for teenagers is often based on coming to terms with the academic impairment rather than aspiring to cure it. However, even in this group, it is imperative to ascertain that knowledge of arithmetic concepts required for daily life, such as understanding a street address, reading calendars, use of money, and writing a check, has been adequately learned.

The Role of the Physician

Clinicians who provide comprehensive care should be aware that developmental dyscalculia is not only a common learning disability but also the most frequently encountered cognitive problem in a variety of medical illnesses. This frequency is the case for children with epilepsy [57], for children with genetic disorders of the X chromosome (i.e., Turner syndrome) [58], and for females with the fragile X syndrome [59]. Developmental dyscalculia also occurs in the context of other developmental cognitive disorders, such as developmental dysphasia, ADHD, and developmental dyslexia [60]. In fact, 26% of kindergarten children with developmental language disorders will manifest significantly impaired arithmetic skills [60], and over 20% of males with ADHD have developmental dyscalculia [61]. Children with ADHD make numerous mistakes in arithmetic, possibly a result of impaired recall, careless errors, inattention to detail, and overall slowness [62]. We therefore advise that before giving a child with ADHD the diagnosis of dyscalculia, the ADHD be adequately treated and arithmetic skills subsequently reevaluated.

For some children with developmental dyscalculia the arithmetic problem is the presenting symptom of a different neurologic illness [63]. In a study assessing children with dyscalculia who were not responding as expected to educational interventions the dyscalculia was found to be the presenting symptom of true absence, ADHD of the inattentive type, developmental Gerstmann’s syndrome, and developmental dyslexia.

We would like to emphasize that the physician’s role in the management of learning disabilities extends beyond the diagnostic phase. This is because physicians, with their medical authority, are often the most appropriate member of the multidisciplinary team to discuss with parents the nature and ramifications of the cognitive disability affecting the child. The physician can provide explanations regarding the neurologic underpinnings of the disorder and its familial-genetic implications, as well as guide both child and parents through the maze of expert assessments. Last but not least, a very important task for which the physician is particularly suited is to aid families in choosing among the plethora of therapeutic modalities available while skirting options best avoided.

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