Number development and developmental dyscalculia

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There is a growing consensus that the neuropsychological underpinnings of developmental dyscalculia (DD) are a genetically determined disorder of ‘number sense’, a term denoting the ability to represent and manipulate numerical magnitude nonverbally on an internal number line. However, this spatially-oriented number line develops during elementary school and requires additional cognitive components including working memory and number symbolization (language). Thus, there may be children with familial-genetic DD with deficits limited to number sense and others with DD and comorbidities such as language delay, dyslexia, or attention-deficit–hyperactivity disorder. This duality is supported by epidemiological data indicating that two-thirds of children with DD have comorbid conditions while one-third have pure DD. Clinically, they differ according to their profile of arithmetic difficulties. fMRI studies indicate that parietal areas (important for number functions), and frontal regions (dominant for executive working memory and attention functions), are under-activated in children with DD. A four-step developmental model that allows prediction of different pathways for DD is presented. The core-system representation of numerical magnitude (cardinality; step 1) provides the meaning of ‘number’, a precondition to acquiring linguistic (step 2), and Arabic (step 3) number symbols, while a growing working memory enables neuroplastic development of an expanding mental number line during school years (step 4). Therapeutic and educational interventions can be drawn from this model.

Developmental dyscalculia (DD) is a specific learning disability affecting the normal acquisition of arithmetic skills. Genetic, neurobiological, and epidemiological evidence indicates that DD, like other specific learning disabilities, is a brain-based disorder, although poor teaching and environmental deprivation have also been implicated in its aetiology.1 However, the developmental course and the interaction of contributing aetiological factors are yet to be clarified. There seems to be a growing consensus that the neuropsychological basis of DD is a disorder of the notion termed ‘number sense’, characterized by deficits in very basic numerical skills such as number comparison.2–5 The term ‘number sense’, introduced by Dehaene,6,7 represents the universal ability to represent and manipulate numerical magnitudes nonverbally on a spatially oriented mental number line. Results from functional brain imaging in adults and in children indicate that the biological equivalent of this ability is a neural network located within the intraparietal sulcus (IPS).8 Number sense is considered to be genetically determined as infants of only a few months of age are able to discriminate exactly (subitize) small and to fuzzy discriminate (approximate) larger sets of objects. These early available preverbal abilities have also been called the ‘core-systems of number’.9

In the present article we argue that the early preverbal core-system abilities are not identical to what school-aged children and adults utilize when operating on a mental number line (see also Giaquinto10). Instead, we argue that the mental number line is a product of experience-dependent, neuroplastic development that requires more than just the availability of intact core-systems, and takes place during preschool and primary school years.11,12 The role of visual imagery, language, and working memory functions has recently been identified as being important in the development of the mental number line.13,14

In the next two sections we will review some current results from neuropsychological and functional brain imaging as well
as clinical and epidemiological research and present a four-step developmental model that allows prediction of different pathways for the subtypes of DD. According to this model, early preverbal core-system representation of cardinal magnitude provides the numerical meaning to number words and Arabic number symbols (digits). Intact counting skills are important for the development of numerical concepts in preschool children; children with specific language impairment may demonstrate impaired acquisition of mathematical skills.\textsuperscript{15,16} To construct, automatize, and successively enlarge a spatial image of ordinal numbers (mental number line), the child needs to interlink the understanding of magnitude (core systems) with the symbolic and spatial-ordinal properties of number. This process requires the cognitive functions which develop during the preschool and primary school years and include, among others, language skills and working memory.\textsuperscript{17}

Therefore, the development of a mental number line that enables cardinal and ordinal processing in higher-order mathematical reasoning may be disturbed in children with DD by reasons of genetic core-system deficits, regardless of comorbidities. However, the presence of comorbidities and developmental issues also needs to be considered as the interactions within the brain, and between the brain and environment, will have an impact on the ultimate phenotype of cognitive development in general and for numbers in particular.\textsuperscript{18}

**Developmental dyscalculia: single or diverse phenomenon**

**RESULTS FROM NEUROPSYCHOLOGY AND FUNCTIONAL BRAIN IMAGING**

One of the most valid experimental indicators for the existence of a spatial-oriented mental number line is the so-called spatial numerical association of response codes (SNARC) effect. The SNARC effect reflects the observation that people are faster to make a judgment about a number if the hand they use to respond is congruous with the size of the number in question: the left hand faster for smaller numbers and the right hand quicker for larger numbers. This effect supports the notion of a mental number line spatially oriented from left (small numbers) to right (larger numbers).\textsuperscript{19} Berch et al. demonstrated that numerical SNARC effects were present in typically developing schoolchildren but not before third grade (7–8y), indicating that this is the time when a left-to-right-oriented mental number line comes into being.\textsuperscript{20}

Schweiter et al. investigated SNARC effects and their possible correlations to number processing and calculation abilities in a sample of children in second grade (6–7y; \( n = 113 \)) from regular primary schools.\textsuperscript{21} A SNARC effect was evident in one-third of the children, but positive correlations between the SNARC effect and mathematical ability could only be demonstrated in male but not female children in second grade. It was argued that the females, although they already possessed a mental number line representation, were not as prepared as the males to use this newly-acquired mental tool to solve mathematical problems. This was attributed to a female preference for language dependent strategies, unlike males who demonstrate a preference for visual-spatial and functional-motor strategies.\textsuperscript{22} With increasing age and expertise, the spatial property of number line representation seems to change according to the amount of its logarithmic compression. Between second and sixth grade (6–7 to 11–12y) children’s numerical estimations between 0 and 1000 become more and more linear.\textsuperscript{11,12}

Results from functional brain imaging research also seem to confirm the experience-dependent aspect of the above hypothesis. Typically achieving children (at age 8–12y) differ from adults as to the amount of neural activity recruited in parietal and prefrontal networks while solving simple arithmetic tasks. Adults typically demonstrate activation primarily in the intraparietal sulcus (IPS), a cortical area that has been proved to be the domicile of the mental number line. Recently, Cohen Kadosh et al.,\textsuperscript{23} using transmagnetic stimulation (TMS), demonstrated the instrumental role of the right hemisphere for automatic magnitude processing. TMS-induced disruption of the right IPS in healthy individuals resulted in the same pattern of responses as in adults who suffer from pure DD.

Children, on the other hand, show significantly lower levels of activity in the IPS arithmetic network but more in frontal regions, areas responsible for attention and executive working memory (i.e. anterior gyrus cinguli).\textsuperscript{24} However, the functional specialization in the parietal cortex for mental arithmetic increases with age and is accompanied by a corresponding decrease of activity in prefrontal regions.\textsuperscript{25,26} This developmental shift, visualized in fMRI studies, provides additional support for the experience-dependent neuroplastic effect on the IPS in the typically developing child.

For children with DD, brain development differs. First, experimental results have shown that the SNARC effect was missing in 7- to 12-year-old children with visual-spatial deficits and DD compared with age-matched controls who did show the SNARC effect.\textsuperscript{27} Furthermore, a recently published fMRI study demonstrated weaker activation than expected in both the parietal and the prefrontal network in children with DD.\textsuperscript{28} This information supports the notion that children with DD suffer from deficits in frontal lobe attentional and executive working-memory resources, in addition to IPS-related visual imagery and mental number line.

**CLINICAL AND EPIDEMIOLOGICAL RESULTS**

DD is a common and heterogeneous phenomenon. Reported prevalence rates from population-based studies range from 4 to 6 per cent\textsuperscript{29} and high rates of different comorbidities, such as attention-deficit–hyperactivity disorder (ADHD), dyslexia, or anxiety disorder,\textsuperscript{30,31} speak against a homogeneous genetic factor hypothesis. The different approaches to the study of DD subtypes have focused either on patterns of strengths and weaknesses within the numerical domain\textsuperscript{32–34} or on patterns of comorbid symptoms, such as dyslexia, ADHD, or visual-spatial and psychomotor functioning deficits.\textsuperscript{25,31,35–37} There may be a subgroup of children with familial-genetic DD with deficits specific to basic numerical domains\textsuperscript{38} and a second group of children with DD and comorbidities.

Using a longitudinal approach, von Aster et al.\textsuperscript{39} investigated early risks, prevalence, and neuropsychology of DD within a representative population-based sample of children (\( n = 578 \)) with a normal estimated nonverbal IQ (>85). The first assessment (T1) took place during the kindergarten year before starting school (mean age 6y 6mo) and the second (T2) at the end of the 2nd grade (mean age 8y 8mo). At preschool level, children were administered the ZAREKI-K (a kindergarten version of the ZAREKI-R [Neuropsychological Test Battery for Number Processing and Calculation in Children – Revised version])\textsuperscript{40} an arithmetic battery covering a broad spectrum of relevant number processing and
calculation abilities. At T2, number processing and calculation abilities were tested with the ZAREKI-R. In addition, reading and spelling skills were measured and teachers completed questionnaires on school performance and behavioural and emotional functioning. The researchers found that the total prevalence of DD was 6%. However, only 1.8% had pure DD (pDD) while as many as 4.2% had comorbid dyslexia (combined dyscalculia and dyslexia [cDD]). The children with cDD had significantly lower scores in ZAREKI-R test performance compared with the pDD group, particularly in those tasks with a high attentional and working memory load. Both groups had similar deficits in tasks of spatial number representations. Not unexpectedly, teacher ratings of ADHD symptoms were significantly higher in the cDD group than the pDD group, the latter scoring at the level of controls.

The total prevalence in this study\(^{39}\) is in accordance with results from other population-based studies\(^{28}\) and supports the findings from Ostad\(^{41}\) and Lewis et al.\(^{42}\) who also found a majority of children with cDD. Furthermore, these results provide evidence for at least two clinically relevant subtypes of DD that can be distinguished by means of comorbidity (dyslexia, ADHD) as well as by the pattern of dysfunctional number processing and calculation abilities. This is in contrast to results reported by Landerl et al.\(^{2}\) that suggest a homogenous dysfunctional pattern in children with pDD and those with cDD, with marked deficits in very basic magnitude comparison tasks for both groups. This may perhaps be attributed to the fact that children with ADHD (a common comorbidity in cDD-children)\(^{39}\) were excluded in that study. Moreover, Rousselle and Noël\(^{43}\) demonstrated that children with DD have more difficulty in assessing number magnitude from Arabic symbols than from non-symbolic numerosity per se, an observation which challenges the single ‘core-system deficit’ hypothesis.

We would argue that the two subgroups of DD differ in their pathways of pathological development. In children with pDD, we speculate that the pathophysiological substrate is mainly an early numerical core-system deficit, probably resulting from a genetic predisposition. In children with cDD, additional or different pathophysiological mechanisms seem operative, including delayed speech and language and impaired attention and executive working memory, causing not only dyslexia or ADHD but also limitations in the development of linguistic, Arabic, and spatial representation of number (mental number line).

There are multiple causes and contributing factors for broad spectrum learning disorders, including obstetric risk factors (e.g. preterm birth or fetal alcohol syndrome), social-environmental deficits and deprivation, early stress-related dysfunctions of emotional regulation and anxiety, as well as genetic dispositions for ADHD or developmental language disorders.\(^{44,45}\)

### A four-step developmental model of number acquisition

We describe a four-step developmental model that enables predictions of possible neuropsychological dysfunctions for DD, with therapeutic modes that can be inferred, recommended, and implemented. Figure 1 illustrates a developmental model of cognitive number representation that is hierarchically organized and could enable prediction of different pathways of pathological development.\(^{46}\) It postulates that the (inherited) core-system representation of cardinal magnitude and accompanying functions, such as subitizing and approximating (step 1), provides the basic meaning of number. This is a necessary precondition for children to learn to associate a perceived number of objects or events with spoken or, later, written and Arabic symbols (\(\bullet \bullet \bullet \rightarrow \) three \(\rightarrow\) 3). The process of linguistic (step 2) and Arabic (step 3) symbolization constitutes in turn a precondition for the development of a mental number line (step 4) in which ordinality is represented as a second (acquired) core principal of number.

If step 1 fails to be established appropriately (i.e. because of genetic vulnerability) names of numbers can be phonologically learned by rote memory, but may function only as non-words, void of the meaning of numerical magnitude. Such children are at risk for pDD. Many of the children with pDD have a neuropsychological profile compatible with Non-verbal

![Figure 1: Four-step-developmental model of numerical cognition. Shaded area below broken line: ‘increasing working memory.’](image-url)
Learning Disability Syndrome\textsuperscript{37} or Developmental Right Hemisphere Syndrome\textsuperscript{48} with marked deficits in visual-spatial and psychomotor functioning.\textsuperscript{33}

If primary core-system abilities are preserved but language development is perturbed, the association between nonverbal numerical properties (\(\bullet\bullet\bullet\)) and their linguistic symbolization (three) cannot be established in an age-appropriate manner (step 2). This could lead to developmental delay in counting routines, counting strategies, arithmetic, and number fact storage. Children with developmental language disorders and dyslexia, therefore, are at risk for cDD. The same may be true for children with primary attentional and working memory deficits. These children have difficulty storing and producing counting sequences or sequences of number word elements (i.e. one hundred and fifty-eight). Inaccuracies and multiple mistakes in arithmetic counting also impede the development of fact retrieval strategies.\textsuperscript{34}

Many children, including those with language and/or ADHD problems, have difficulties acquiring the Arabic notation system in preschool and first years of elementary school with its place value syntax and the corresponding transcoding rules (step 3). This is especially true for children who have to transcode from two or more linguistic number word systems into the Arabic code, when being educated in a foreign language or for bilingual children.\textsuperscript{34} Such children often become confused when confronted with different linguistic irregularities in the different languages which require special rules for transcoding from each language into the Arabic notation and vice-versa.\textsuperscript{39} For example, in ‘twenty-five’ the positions of tens and ones correspond to their positions in the Arabic notation. However, in German, the number word is \textit{funf-und-zwanzig} whereby the ones precede the tens, requiring the child to change positions when transcoding into the Arabic syntax. To ignore that rule leads to a numeral of 52 that will be located on the arising mental number line between 50 and 60. Such erroneous experiences may disturb the construction and automatization of a spatial image of ordinal numbers.

The mental number line (step 4) comes into play when identifying ordinal positions of numbers with reference to their numerical neighbours. Milestones on the mental number line (i.e. tens and hundreds) are marked in Arabic notation and provide orientation when roving mentally during arithmetic thinking. The mental images of the number line have been reported to be quite different among certain individuals and may have a more complex structure than the theoretical straight line.\textsuperscript{50} Different synesthetic associations of numbers (i.e. colour and brightness) also have been described.\textsuperscript{51} This illustrates that the development of cognitive number representations is strongly influenced by individual experiences and learning environment.

Usually in pDD-children early core system abilities and frequently basic visual-spatial processing abilities are impaired. In most cDD-children, who compose the large majority of children with dyscalculia, developmental disorders of attention and executive working memory or speech and language seem to impede the typical acquisition of number concepts. These concepts range from the linguistic and Arabic number system to the construction of the mental number line. In our experience, core system abilities seem to be intact in many of these children (see also \textsuperscript{35}).

However, there is a differential diagnosis. Classical fear conditioning and accompanying reductions of working memory may cause a circumscribed inability to mentally construct within the parietal network a mental number line in otherwise typically developing children. Even in a total absence of any basic neuropsychological dysfunction or comorbid condition, DD may occur as a result of early dysfunctional learning experiences.

\textbf{Case report}

An example of how maladaptive experiences may influence the emergence of DD is the case of Irma:

At second grade, teachers told Irma’s parents that their 8-year-old child had severe dyscalculia, verified by a test of mathematical achievement. Reading and spelling skills had been excellent. Irma developed a deep aversion to mathematics and was absolutely convinced that she would never learn arithmetic. She also showed increasing symptoms of depression and reluctance to attend school. She was, however, able to compare the magnitude of different amounts of objects, indicating that core-system abilities were intact. Irma had above average verbal and nonverbal intelligence and she engaged in rich and imaginative play. At about 3 to 4 years of age Irma started to invent a fantasy play, a kind of fairy tale in which the protagonists, unfortunately, had numbers for names. During therapy sessions she would paint and give detailed biographies of all the persons in her fantasy world: ‘Three’ was a lovely boy, but sometimes cheeky to his mother. He had two friends in his neighbourhood, ‘nine’ and ‘twenty-three’, with whom he did a lot of silly tricks, and so on…

One can easily imagine how confusing it might have been for Irma when she was asked by her teachers to subtract 3 from 9. Irma constructed a semantic number module in early childhood that was rather incompatible with the required numerical meaning and to an image of an ordinal mental number line. She extensively used numbers for non-numerical assignments and associations, which sometimes can also be observed in autistic children. Irma’s case was quite extraordinary, but it demonstrates the importance of a careful exploration of the individual learning history and that a conventional number line isn’t available for every child.

\textbf{Conclusion}

If DD is indeed a disorder of number sense, and number sense is conceptualized as a disorder of early core-system abilities (subitizing, approximation), then the disorder that Irma manifested was not dyscalculia. In turn, if number sense is conceptualized as availability of a mental number line, Irma was dyscalculic. Within the proposed four-step developmental model, number sense is defined by both, an inborn part (step 1: cardinal magnitude) and an acquired part of semantic number representation (step 4: ordinal number line). According to Karmiloff-Smith’s developmental theory,\textsuperscript{52,53} the latter (step 4) can be viewed as a representational redescription of the former (step 1), which requires the intermediate development of domain specific abilities (steps 3 and 4: linguistic and Arabic number representation), as well as domain general capacities (attention, executive working memory). Disorders of number sense, therefore, may have genetic as well as developmental and environmental reasons.
Disorders of attention, emotional and behavioural regulation, and language development lead to comorbid conditions in preschool and school years, and require consideration when planning interventions. Psychotherapy, pharmacotherapy, and specific reading and spelling training should be instituted according to need. Specific training to enhance number processing and calculation abilities should be designed individually, taking into consideration the profile of strengths and weaknesses of each child.

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References

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<td>DD</td>
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<td>cDD</td>
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<td>IPS</td>
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<td>SNARC</td>
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