Co-occurrence of developmental disorders: The case of Developmental Dyscalculia

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Abstract

Five to seven percent of children experience severe difficulties in learning mathematics and/or reading. Current trials that are focused on identifying biological markers suggest that these learning disabilities, known as Developmental Dyscalculia (DD) and Dyslexia (for reading), are due to underlying brain dysfunctions. One ongoing controversy concerns the extent to which arithmetic impairments are specific to DD or shared with other developmental disorders such as Dyslexia. This review explores and develops a hypothesis for cases of DD + Dyslexia. Three factors warrant consideration: (a) the behavioral factor, including definitions of the disabilities and assessment tools; (b) the cognitive factor, including whether co-occurrence of DD and other developmental disorders such as Dyslexia derive from similar or different cognitive risk factors; (c) the biological factor, including consideration of static vs. developmental neuropsychology. Better understanding of the causes of co-occurrence of DD and Dyslexia, or other developmental disorder such as Attention Deficit Hyperactivity Disorder (ADHD), can have an important influence on research that examine the two disorders, including research on therapy and etiology.

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Mathematical skills are fundamental in most societies and have great importance not only to individuals but also for educational and health systems. Depending on the country, current estimates are that 5–6% of children fail to show typical development of numerical abilities. Hence, a significant proportion of the school-age population will have a specific learning deficit in mathematics known as Developmental Dyscalculia (DD).

Here, however, I would like to use the distinction between DD and Mathematical Learning Disabilities (MLD) made by Rubinsten and Henik (2009). Both are disorders in mathematics with no other non-numerical disorder. The term DD is reserved here for a deficit in core numerical abilities (e.g., difficulty in processing quantities). By contrast, MLD is the result of several general cognitive deficits such as deficient working memory, visual-spatial processing or attention.

The last decades of the 20th century saw the development of ideas linking cognitive psychology, education and neuroscience and major strides have been made toward understanding the brain mechanisms involved in arithmetic reasoning. One of the results of these developments is that DD has undergone transformation from a rather debatable term into a stimulating issue; and rightly so, given its prevalence of 5–6% (von Aster & Shalev, 2007; Wilson & Dehaene, 2007). However, developments in cognitive neuroscience have led to a conception that this disorder is caused by a discrete cognitive deficit, independent of other cognitive abilities. Specifically, it is well established that children, adults, and nonhuman animals have the innate capacity to perceive and discriminate small numerical quantities (for review see Ansari, 2008; Cantlon, Platt, & Brannon, 2009). Current trials focused on identifying reliable biological markers suggest that DD is due to underlying brain dysfunctions (Price, Holloway, Vesterinen, Rasanen, & Ansari, 2007) known to be involved in the processing of quantities (Kaufmann et al., 2005): the Horizontal Intra Parietal Sulcus (HIPS—Cappelletti, Barth, Fregni, Spelke, & Pascual-Leone, 2007; Cohen Kadosh et al., 2007; Kucian et al., 2006; Mussolin et al., in press; Price et al., 2007; Rotzer et al., 2008). Accordingly, DD is currently understood to reflect deficient numerosity understanding (i.e., intuitions for quantities resulting in mental representation of quantities or magnitudes; Butterworth, 2005; Iuculano, Tang, Hall, & Butterworth, 2008; Landerl, Bevan, & Butterworth, 2004; Rousselle & Noel, 2007; Wilson & Dehaene, 2007). However, behavioral deficits seen in DD are heterogeneous (Dowker, 2008). Also, as in the case of many other developmental disorders (Gilger & Kaplan, 2001; Karmiloff-Smith, 2009), multiple problems are the rule; pure disorders apply only to a minority of cases (Kaufmann & Nuerk, 2005; Rubinsten & Henik, 2009). Specifically, studies show that 20–60% of children with DD have associated learning problems such as Dyslexia (Dirks, Spyer, van Lieshout, & de Sonneville, 2008; Mayes & Calhoun, 2006) or Attention Deficit/Hyperactivity Disorder (ADHD; Capano, Minden, Chen, Schachar, & Ickowicz, 2008; von Aster & Shalev, 2007). For example, research of a German sample of children (n = 378) found that although the prevalence of dyscalculia is about 6%, only 1.8% had pure dyscalculia, while 4.2% had co-morbid dyslexia (von Aster, Schweiter, & Weinhold Zulauf, 2007).

Accordingly, an ongoing controversy concerns the extent to which arithmetic impairments are specific to DD or shared with another developmental disorders such as Dyslexia (Simmons & Singleton, 2008; Tressoldi, Rosati, & Lucangeli, 2007; von Aster & Shalev, 2007). Based on Rutter and Sroufe (2000) and Morton and Frith (1995), it is suggested here that three factors should be considered in order to address this issue. One is the behavioral factor, including definitions of the disabilities and the use of assessment tools. Specifically, it is possible that some cases of co-morbidity are driven from using wrong diagnostic criteria and processes. A second is the cognitive factor. Both co-occurring DD and other developmental disorders such as Dyslexia may derive from intercorrelated cognitive risk factors. On the other hand, the presence of one form of developmental disorder may, through its effects, constitute a risk mechanism for another form of developmental disorder (Rutter & Sroufe, 2000). A third is the biological factor. Consideration of the static vs. developmental neuropsychology should be taken into account. Specifically, the static adult neuropsychological model may be unsuitable for the study of developmental disorders (Karmiloff-Smith, 2006, 2009) such as DD.
The aim of this article is to examine how specific cognitive functions promote DD and MLD. Guided by the argument that co-morbidity creates challenges for characterizing disorders and their causes (Angold, 1999; Caron & Rutter, 1991), I propose several hypotheses with respect to behavioral, cognitive and biological factors in cases of pure vs. co-morbid developmental disorders. Therefore, this work reviews not only DD but also co-occurring DD and other developmental disorders. The focus here is on DD + Dyslexia or MLD + Dyslexia but due to limited research in the field, cases of DD + ADHD are discussed as well.

1. Factors to be taken into account in DD

1.1. The behavioral factor: diagnostic criteria and tools

Just as DD has the HIPS as a biological marker, Dyslexia is also considered a brain-based disorder (that involves unusual patterns of brain function). Dyslexia is a familial, persistent, and impairing specific learning disability, characterized by unexpected difficulties with accurate and/or fluent word recognition, poor spelling and decoding abilities, none of which can be ascribed to sensory difficulties, and possibly low IQ or inadequate education (Directors, 2002). Brain imaging studies have largely confirmed structural and functional abnormalities in occipitotemporal brain areas (termed the Visual Word Form Area; Cohen et al., 2000) in many cases of Dyslexia (Démonet, Taylor, & Chaix, 2004; Eckert, 2004; Gaillard et al., 2006; McCandliss & Noble, 2003; Shaywitz & Shaywitz, 2008).

Nevertheless, neither DD, MLD nor Dyslexia can currently be diagnosed by biological markers. Instead, the diagnosis is based on behavioral criteria (Siegel, 2006; von Aster & Shalev, 2007). However, we know little about how behavioral aspects of MLD vary as a function of co-morbidity. This is so because there has been a markedly small number of both neuro-cognitive and behavioral studies (Rourke & Finlayson, 1978; Shalev, Manor, & Gross-Tsur, 1997). Most of this work is based on paper-and-pencil behavioral tests that involve higher-level cognitive processes in addition to the basic numerical processes that are of interest (Ansari & Karmiloff-Smith, 2002). For example, Tressoldi et al. (2007) tested two children with Dyslexia only, two with DD only, and three more with DD + Dyslexia. Results indicated that deficits in numerical abilities, such as mental and written calculations, arithmetical facts retrieval, number comparison and number alignment were not associated with Dyslexia.

Contrary to Tressoldi et al., Jordan, Hanich, and Kaplan (2003) found that students at the end of third grade both with MLD only and MLD + Dyslexia perform equally on arithmetic fact retrieval, approximate arithmetic and place value. Jordan et al. presented results suggesting that MLD and MLD + Dyslexia are co-morbidly associated (i.e., the math problem is the same type of disorder in both MLD and MLD + Dyslexia). Robinson, Menchetti, and Torgesen (2002) propose a way to resolve this contradiction. They suggest that deficiencies with arithmetic fact retrieval are associated with MLD but only with some cases of MLD + Dyslexia.

However, Hanich (2001) showed that in mid second grade, MLD children are better than MLD + Dyslexia on exact calculations and story problems. At the end of third grade (Jordan et al., 2003) the MLD group is better than the MLD + Dyslexia group only on calculation principles. Also, Jordan, Kaplan, and Hanich (2002) found that in early elementary school, children with MLD only progress faster in mathematics achievement than do children with co-morbid MLD + Dyslexia. Finally, phonological processes (i.e., phonological memory, rate of access to phonological information, and phonological awareness) were found to contribute significantly to the development of computational skills between second and fifth grade and to account for a large part, if not all, of the association between MLD and Dyslexia in cases of MLD + Dyslexia. This may suggest that phonological processing deficits of individuals with Dyslexia (with or without MLD) impair aspects of mathematics such as arithmetic facts retrieval (Simmons & Singleton, 2008). These findings also may reflect the likelihood that MLD and MLD + Dyslexia are not co-morbidly associated (i.e., the math problem is not the same type of disorder in both MLD and MLD + Dyslexia) but Dyslexia and MLD + Dyslexia are indeed co-morbidly associated.

It is clear that the resolution of the debate concerning the extent to which arithmetic impairments are specific to DD or shared with Dyslexia is challenged by the marked heterogeneity in behavioral symptoms in both Dyslexia and DD (Simmons & Singleton, 2008; Tressoldi et al., 2007; von Aster &
In the last two decades, researchers have made impressive strides in studying numerical cognition and brain mechanisms involved in DD by using low-level cognitive tasks instead or in addition to paper-and-pencil tasks. Recently, for example, a double dissociation between DD and Dyslexia was found, in the ability to automatically associate quantities with written numbers vs. automatically associating phonemes with written letters (Rubinsten & Henik, 2006). However, to our knowledge, only two such low-level studies have addressed co-morbidity issues. Landerl et al. (2004) argued that children with DD and DD + Dyslexia do not have a different type of deficit but suffer from the same number-processing deficit. Both groups of children with DD only and DD + Dyslexia were slower on tests that included counting dots, comparing values of single digits, reciting number sequences, reading three-digit numbers and writing numbers. Also, Rousselle and Noel (2007) found no evidence for differential patterns of performance between children with DD and DD + Dyslexia in tasks assessing basic numerical skills. Their argument states that the deficit in both DD and DD + Dyslexia is in the ability to associate quantities with written numbers.

Estimates of DD + Dyslexia vary not only due to behavioral assessment tools but also due to diagnostic criteria. For instance, one study reports combined reading and arithmetic problems among 7.6% of school-aged children using a cut-off score of below the 25th percentile, but 1% of school-aged children using scores below the 10th percentile (Dirks et al., 2008; Ramaa & Gowramma, 2002). Indeed, diagnosis of DD is generally determined psychometrically on the basis of low scores on individually administered standardized tests of arithmetic achievement, but there is no consensus as to the kind of standardized tests and the threshold score to be used in decision making (Mazzocco & Myers, 2003). This fact highlights the need to distinguish between MLD and DD partly based on different threshold scores in standardized tests (Murphy, Mazzocco, Hanich, & Early, 2007; Rubinsten, Bedard, & Tannock, 2008).

To summarize, both DD and Dyslexia can be defined as neuro-developmental disorders with a biological origin that possess specific behavioral signs (Cappelletti et al., 2007; Cohen Kadosh et al., 2007; Kucian et al., 2006; Mussolin et al., in press; Price et al., 2007; Rotzer et al., 2008; Shaywitz & Shaywitz, 2008). These signs expand considerably beyond mathematical problems (which are behavioral symptoms). Therefore, some causes of the behavioral signs, as well as symptoms of the condition, can, and perhaps should be, specified cognitively. For example, the ability to automatically associate written symbols with mental representations such as quantities or phonemes may lead to math and reading difficulties. These theoretical deficits are not conclusively proven scientifically, but they can serve as a basis for testable predictions at both the behavioral and the biological levels. In other words, the core features of these two disorders may be best understood in terms of deficits at the cognitive level (Frith, 2001). Therefore, when cognitive factors are measured, and not only behavioral symptoms, an easier and more precise diagnosis of DD only vs. DD + Dyslexia can be attained.

1.2. The cognitive factor: single vs. multiple cognitive risk factor

Co-morbidities of either DD or MLD together with other developmental disorders could be due to several different cognitive deficits or a single cognitive deficit that through its effects becomes a risk mechanism for another form of developmental disorder. An example of this phenomenon can be seen in the field of ADHD. A substantial proportion of individuals with ADHD manifest unexpected problems in mathematics that cause an impairment in academic achievement and daily functioning, with estimates ranging from 10% to 60% (Capano et al., 2008; Mayes, Calhoun, & Crowell, 2000). However, in contrast to co-occurring reading difficulties, the nature of mathematical difficulty in ADHD has received little attention from teachers or researchers. Some researchers attribute the significant mathematical delays in children with ADHD to attention-based impairments (Lindsay, Tomazic, Levine, & Accardo, 2001) or working memory (Rosselli, Matute, Pinto, & Ardila, 2006). These general cognitive impairments (i.e., not specific to mathematics) are considered to be integral features of the ADHD syndrome and, hence, may cause mathematical difficulties in some of these children (i.e., MLD + ADHD) (Castellanos, Sonuga-Barke, Milham, & Tannock, 2006). An alternative proposition is that subgroups of children with ADHD and mathematical difficulties may exhibit different underlying mechanisms, including specific deficits in basic numerical processing (e.g., quantity processing), as manifest in children with Developmental Dyscalculia (i.e., two major cognitive deficits resulting in DD + ADHD).
Rubinsten et al. (2008) investigated effects of stimulant medication (methylphenidate; MPH) on arithmetic performance in children with ADHD. They identified three groups of children with ADHD from an existing large database: one group with DD (DD + ADHD), one group with more general and less severe difficulties in arithmetic (MLD + ADHD), and one group with good arithmetic abilities (ADHD). Children with DD + ADHD exhibited both general cognitive dysfunctions and specific deficits in understanding quantities. In contrast, arithmetic difficulties in children with MLD + ADHD were associated with deficits in executive function and working memory. Furthermore, MPH enhanced performance in arithmetic problems dependent upon working memory (involving activation in the frontal lobes), but not upon processing numerical quantity (involving activation in the parietal lobes). These findings suggest the importance of distinguishing between DD + ADHD and MLD + ADHD. Namely, in some children, ADHD impairments in attention or working memory also cause mathematical difficulties resulting in co-occurrence of MLD. However, cases of DD + ADHD are the result of double deficits in both quantity processing and executive functions. Kaufmann et al. have found that some 9- to 12-year-old children with ADHD (but no other developmental disorder) have deficient numerical processing skills, specifically in their ability to process numerical magnitude (Kaufmann & Nuerk, 2008).

1.3. The biological factor: static neuropsychological vs. developmental approaches

Most current diagnostic criteria are based on a static neuropsychological approach similar to that used in the case of adults who acquire brain damage later in life. In such cases, a direct connection exists between damage to a specific brain area and a specific cognitive deficit such as quantity processing. Accordingly, an acceptable assumption is that DD only, Dyslexia only and even DD + Dyslexia are each discrete developmental disorders with clear boundaries.

For example, studies show that the angular gyrus (a brain area in the parietal–temporal lobe, located posterior and inferior to the HIPS) is involved with both reading and math and, hence, dysfunctions in this brain area may cause both DD and Dyslexia to different extents. Specifically, the angular gyrus is considered to be involved in reading (see Shaywitz & Shaywitz, 2008, for discussion of the importance of the parietotemporal reading system in developmental dyslexia) and shows increased activation in response to structured phonological intervention programs (Shaywitz et al., 2004). The angular gyrus is also considered to be part of the numerical network that is activated during exact calculation (García-Orza, León-Carrión, & Vega., 2003; Grabner et al., 2009; Van Harskamp, Rudge, & Cipolotti, 2002) and involves the left inferior frontal lobe and bilateral angular gyri (Dehaene, Piazza, Pinel, & Cohen, 2003). For example, repetitive transcranial magnetic stimulation (rTMS) over the left angular gyrus disrupted number magnitude processes (Rusconi, Walsh, & Butterworth, 2005). Also, an intracranial electro-stimulation study identified areas within the left angular gyrus that were specifically involved in either multiplication or subtraction (Duffau et al., 2002). Hence, from a neuropsychological perspective, a deficit in the angular gyrus may cause both reading impairments (Horwitz, Rumsey, & Donohue, 1998) and calculation difficulties (DD) (Grabner et al., 2009).

To better understand DD and co-occurring of other developmental disorders, however, developmental calculation models need to be distinct from adult calculation models (Karmiloff-Smith, 2006, 2009; Van Herwegen, Ansari, Elsabbagh, Xu, & Karmiloff-Smith, 2008). Accordingly, a different biological working hypothesis can be suggested. Instead of being genetically pre-specified, the involvement of the angular gyrus with cognitive functions develops through interaction with the environment. In this case, deficiencies in several different cortical pathways and brain areas that are involved with the most basic numeric and phonological processes and are functionally connected to the angular gyrus (e.g., IPS and parietal–temporal brain areas) may harm the specialization in the angular gyrus through development and training. Namely, deficient firing from the IPS (involved with quantity processing and hence with DD) and from the temporal–parietal brain areas (involved with phonological processing and hence, with Dyslexia), may lead to deficient links between each of these areas and the angular gyrus at the end of development and training. This may result in DD + Dyslexia.
The suggestion of multiple cognitive deficits (e.g., processing quantities, phonemic awareness and associating symbols with quantities or with phonemes) due to multiple brain dysfunctions (e.g., IPS, temporal–parietal brain areas and angular, respectively) fits better with a developmental approach. For example, studies employing short-term training of arithmetic problems in adults have demonstrated training–related changes in parietal regions that are typically involved with calculation (Delazer et al., 2003, 2005; Ischebeck, Zamarain, Egger, Schocke, & Delazer, 2007; Ischebeck et al., 2006). Delazer et al. trained typically developing participants on complex multiplication problems. After training, in an fMRI test meeting they found a shift of activation within the parietal lobe from the intraparietal sulcus to the left angular gyrus, suggesting a change from quantity-based processing to verbal retrieval from memory. Accordingly, behavioral symptomatology consistent with DD + Dyslexia could be the result of atypical development affecting neural connections between several relevant brain areas (Johnson, 2001).

Furthermore, such a developmental approach is consistent with arguments suggesting that expression of genes in the neocortex tends to extend to most brain regions and that a specific gene involved with a very specific brain region, has not yet been found (Karmiloff-Smith, 2006). Accordingly, deficiencies in several different cortical pathways that are functionally connected to each other may be genetically caused and/or created through development and training. This suggests that co-occurring DD and Dyslexia may derive from a set of intercorrelated risk factors.

2. Summary and conclusions

This article has been concerned mainly with exploring and developing a hypothesis for cases of DD + Dyslexia. It is important to note that the hypothesis only suggests empirical alternatives without constraining the researcher. For example, the angular gyrus is considered here as a possible brain area that, in addition to the IPS, is involved in DD + Dyslexia. However, it is offered only as a suggestion. Another innovative hypothesis is involvement of the primary visual brain area in cases of DD + Dyslexia. This hypothesis is based on recent evidence showing that this brain region is involved in estimating quantities (Burr & Ross, 2008). Also, recent work suggests that left-lateralized white matter structures (specifically the superior corona radiate) are involved with both reading abilities and mathematical competence and hence, may be associated with DD + Dyslexia (Ben-Shachar, Dougherty, & Wandell, 2007; van Eimeren, Niogi, McCandliss, Holloway, & Ansari, 2008).

The two subgroups DD and Dyslexia should probably be attributed to different underlying cognitive problems that most likely implicate dysfunction of different neural networks. However, in some cases, malfunctioning of a unique cognitive function, such as working memory, could produce different pathologies presented, for example, as MLD + ADHD (Rubinsten et al., 2008). Accordingly, at the behavioral and cognitive levels, when studying co-morbidity, researchers should consider multiple phenotypes. A limited phenotype may miss much of the complexity of an individual’s atypical development (Gilger & Kaplan, 2001).

Also, very little is known about the molecular biological origins of DD or MLD, and there are very few longitudinal studies that examine developmental aspects of these disorders. I emphasize brain dysfunction as a possible biological origin. However, DD or MLD may involve genetic or environmental factors.

Better understanding of the causes of co-morbidity between MLD and Dyslexia, or another developmental disorder such as ADHD, can have an important influence on future research that examine the two disorders. This includes research on therapy and etiology (Frith, 2001). For example, if there is indication that people with both MLD and Dyslexia, actually have a third, distinct disorder with a different etiology, it is likely that a treatment that is efficient for one of the disorders when it occurs alone may not be efficient for people with both disorders (Rubinsten et al., 2008). These possibilities warrant further investigation.

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