To appear in: Human Behavior, Learning, and the Developing Brain: Atypical Development. Edited by Coch D, Dawson G, Fischer K: Guilford Press, New York.

RUNNING HEAD: Number Sense and Developmental Dyscalculia

Number Sense and Developmental Dyscalculia

Anna J. Wilson and Stanislas Dehaene

INSERM-CEA Unit 562 « Cognitive Neuroimaging »

Service Hospitalier Frédéric Joliot

CEA-DRM-DSV

Orsay, France

Abstract

In this chapter we review the possible biological bases for developmental dyscalculia, which is a disorder in mathematical abilities presumed to be due to impaired brain function. By reviewing what is known about the localization of numerical cognition functions in the adult brain, the causes of acquired dyscalculia, and the normal development of numerical cognition, we propose several hypotheses for causes of developmental dyscalculia, including that of a core deficit of "number sense" related to an impairment in the horizontal intra-parietal sulcus (HIPS) area. We then discuss research on dyscalculia, including the contribution of recent imaging results in special populations, and evaluate to what extent this research supports our hypotheses. We conclude that there is promising preliminary evidence for a core deficit of number sense in dyscalculia, but we also emphasize that more research is needed to test the hypothesis of multiple types of dyscalculia, particularly in the area of dyscalculia subtyping. We complete the chapter with a discussion of future directions to be taken, the implications for education, and the construction of number sense remediation software in our laboratory.

Introduction

Claire is an 8 year old second grader who participated in a remediation study for dyscalculia which we have recently conducted. Despite an average IQ and good motivation and attention, she struggles in school, especially in math. This does not seem to be due to her reading abilities; her reading speed is average, and although she has some trouble with comprehension, she already receives special education services for this, with no seeming improvement in her mathematical performance. Our tests confirm that her basic numerical skills are far behind those of her peers: She shows a developmental lag in counting, understanding of place value, and in addition and subtraction of one digit numbers. The latter are only around 80 and 40 percent accurate respectively, and both are carried out in a painstaking fashion using finger counting.

Claire is a fairly typical case of developmental dyscalculia, which is generally defined as a disorder in mathematical abilities presumed to be due to a specific impairment in brain function (Kosc, 1974; Shalev & Gross-Tsur, 1993, 2001). This definition is highly similar to that of "Mathematics Disorder" in the DSM-IV (American Psychiatric Association, 1994), and also that of "mathematical learning disabilities" (Geary, 1993, 2004). Because of this, and following Butterworth (2005a, 2005b), we will take these constructs to be one and the same[i].

Despite its professional and practical consequences (e.g. Rivera-Batiz, 1992), and a similar population prevalence of around 3-6% (Badian, 1983; Gross-Tsur, Manor, & Shalev, 1996; Kosc, 1974; Lewis, Hitch, & Walker, 1994), developmental dyscalculia is much less recognized, researched and treated than its cousin developmental dyslexia. This has partially been a consequence of the later development of our knowledge about the neural bases of numerical cognition, as opposed to reading. As research in the numerical cognition field has started to increase in volume, so has cognitive neuroscience research on dyscalculia, with several research teams conducting studies on its associated cognitive profile, brain bases, and genetics.

The core deficit hypothesis

An advantage provided by the good twenty or so year lag between dyscalculia and dyslexia research is an opportunity to speed up research on the former by looking for analogies that may be made between the two disorders. One interesting and important analogy that has been made, for which we will review evidence in this chapter, is that of a "core deficit". The core phonological deficit hypothesis is now accepted by many in the dyslexia field (Goswami, 2003), and has resulted in many advances in prevention and remediation (Eden, 2002; Tallal et al., 1996; Temple et al., 2003). Importantly, the nature of the core deficit can be somewhat counter-intuitive – who would have thought that training children in distinction of sounds would improve reading? However, this idea is now confirmed by our knowledge of the brain circuits involved in reading, especially the continuing role of phonological areas in reading in even practiced readers.

Could there be a similar core deficit in dyscalculia? Although our knowledge of its behavioral manifestations is incomplete, our knowledge of the adult circuits involved in numerical cognition is by now fairly advanced. It has been argued that the core aspect of numerical cognition is "number sense", which is a short-hand term for our ability to quickly understand, approximate and manipulate numerical quantities (Dehaene, 1997, 2001). We now have a plausible candidate for a neural substrate of number sense: a specific region of the parietal cortex, the horizontal intra-parietal sulcus (HIPS), which based on neuroimaging results is hypothesized to contain a non-verbal representation of numerical quantity, analogous to a spatial map or "number line" (Dehaene, Piazza, Pinel, & Cohen, 2003).

Our knowledge of the intact adult system allows us, therefore, to make a prediction about the impaired system in the child. The behavioral hypothesis that a deficit in number sense is the cause of at least some types of dyscalculia has in fact already been previously proposed by authors in the special education field (Gersten & Chard, 1999; Robinson, Menchetti, & Torgesen, 2002) and in the numerical cognition field (Butterworth, 1999). Here we propose a neural specification of this hypothesis: that at least some types of dyscalculia may be due to an impairment of functioning and/or structure in the HIPS, and/or in its connections to other numerical cognition regions. In this chapter we will examine how much evidence there is to support this hypothesis.

A complementary hypothesis: multiple subtypes of dyscalculia

As well as presenting and discussing the hypothesis of a "core deficit", we also present and discuss the possibility of multiple causes of dyscalculia. Although a majority of children might suffer from a core impairment in number sense, other sources of dyscalculia are likely to exist. Hypotheses about their nature may be generated from several sources, such as the adult neuroimaging literature, the developmental literature, and educational research on subtypes of dyscalculia. To take one example, in the adult neurological literature it is possible to find a "number sense" acalculia, in which the patient has lost all sense of the meaning of numbers, but it is also possible to find a "number fact retrieval" acalculia, in which the patient still understands the meaning of numbers, but is unable to retrieve from memory basic multiplication or addition facts, and is thus forced to laboriously recalculate these facts each time by counting, or worse retrieves the wrong result without noticing (Dehaene & Cohen, 1997). We will also discuss whether there is evidence for a subtype of developmental dyscalculia similar to this latter type of patient.

Outline of the chapter

We start by describing cases of adult acquired dyscalculia, and discussing what we know about the underlying numerical cognition systems in the adult. We then overview the developmental numerical cognition literature. The combination of these two literatures allows us to elaborate some predictions about possible types and causes of developmental dyscalculia. Next we discuss in depth developmental dyscalculia: how it is identified, its characteristics and common comorbid disorders, and what we have learnt from subtype research. We then discuss the extent to which this literature supports our hypotheses, in particular the behavioral and neural evidence for a deficit in number sense. Finally we discuss future directions to be taken, the implications for education, and the construction of number sense remediation software in our laboratory.

Adult numerical cognition

There are two important areas of adult numerical cognition research which can be used to shed light on the possible causes of developmental dyscalculia. The first is neuropsychological research on acquired dyscalculia (referred to here as "acalculia"), which may have similar causes and symptoms to developmental dyscalculia (referred to here as "dyscalculia"). The second is our knowledge of the functioning of the intact numerical cognition circuits in the adult brain, which gives us a clue as to what kind of deficits we might expect if developmental dyscalculia is caused by abnormalities in these circuits.

Acquired dyscalculia or "acalculia": lesion evidence

The adult neuropsychological literature supports a causative role of inferior parietal lesions in acalculia (see Cohen, Wilson, Izard, & Dehaene, in press, for a more detailed review of this literature). Acalculia is often associated with Gerstmann's syndrome, in which patients also present left-right disorientation, agraphia, and finger agnosia (Gerstmann, 1940). This syndrome is usually (although not always) observed due to lesions around the region of the left angular gyrus (Jackson & Warrington, 1986; Rosselli & Ardila, 1989). However, the existence of a coherent syndrome has been questioned, as it appears that the four deficits are able to be dissociated (Benton, 1992).

Dehaene and Cohen (1997) discuss two cases of acalculia (patients MAR and BOO) who together provide a double dissociation between a "number sense" type acalculia and a "verbal memory" type acalculia. Patient MAR, a left-hander with a right inferior parietal lesion and a pure Gerstmann's syndrome, showed difficulties with tasks requiring quantity manipulation, such as choosing the larger of two digits, bisecting number lines, and subtraction. Patient BOO, a right-hander with a left subcortical lesion showed a very different pattern, with few difficulties in the former tasks, but severe difficulties with more verbal or memory-related numerical tasks, such as multiplication. Other similar cases have also been reported more recently by Lemer et al. (2003), Capeletti et al. (2001) and Delazer et al. (in press).

To the extent that acquired and developmental dyscalculia are similar in their causes and manifestations, the acalculia literature thus suggests two important hypotheses for developmental dyscalculia: a) that we should expect to find more than one type of dyscalculia, in particularly we should expect to find a "number sense" dyscalculia, and a "verbal memory" dyscalculia, and b) that we should expect to find a link between "number sense" dyscalculia and the inferior parietal lobes, particularly the angular gyrus. In the next section we will see that the adult imaging literature supports these two suggestions, whilst adding further precision to our hypothesis about anatomical location.

Imaging evidence: number sense and the horizontal intra-parietal sulcus

Early PET imaging studies consistently showed activation in the parietal cortex during numerical tasks (Dehaene et al., 1996; Pesenti, Thioux, Seron, & De Volder, 2000; Zago et al., 2001), although only some of these early results (e.g. Dehaene et al., 1996) supported a dissociation between areas activated by quantity manipulation vs. rote memory tasks, as would be expected from the neuropsychological literature.

Later fMRI studies provided much more precision, and in a recent meta-analysis of available data from this literature, Dehaene et al. (2003) put forward a more precise localization hypothesis, identifying three particular areas of the parietal lobe which appear to be differentially involved in representation and processing of numerical information (See Figure 1). The left angular gyrus, the classic Gerstmann's lesion center, appears to be more active in more verbal numerical tasks such as multiplication and exact addition (Chochon, Cohen, van de Moortele, & Dehaene, 1999; Dehaene, Spelke, Pinel, Stanescu, & Tsivkin, 1999; Lee, 2000). The posterior superior parietal lobe (PSPL) appears to be activated in numerical tasks which may require the shifting of spatial attention, such as approximating, subtraction and number comparison (Dehaene et al., 1999; Lee, 2000; Pinel, Dehaene, Riviere, & LeBihan, 2001).

*** INSERT FIGURE 1 AROUND HERE ***

In contrast, the nearby horizontal intra-parietal sulcus (or HIPS) appears to be more active in core quantity manipulation or "number sense" tasks such as comparing the size of numbers, estimating, subtracting, and approximating (Chochon *et al.*, 1999; Dehaene *et al.*, 1999; Lee, 2000). This role of the HIPS in quantity representation has been further reinforced by subsequent imaging studies (Eger, Sterzer, Russ, Giraud, & Kleinschmidt, 2003; Pinel, Piazza, Le Bihan, & Dehaene, 2004), and by the recent finding by Piazza et al. (2004), using an fMRI habituation paradigm, that fMRI adaptation in this area shows tuning curves similar to those found in single neuron recordings from the analogous area of the monkey brain (Nieder & Miller, 2004).

Overall, the neuroimaging literature leads to the same hypotheses arrived at from the neuropsychological literature: we should expect to find a role of the inferior parietal lobes in dyscalculia, and we should be able to find different subtypes of dyscalculia. These subtypes should be a "number sense" dyscalculia linked to impairment (functional or structural) of the HIPS, a "verbal" dyscalculia linked to impairment of the angular gyrus, and a "spatial attention" dyscalculia linked to impairment of the PSPL.

However one caveat is that these "theoretical subtypes" might be quite difficult to dissociate in developmental cases. Firstly, in a developmental context, any deficit in one of the numerical systems is likely to interfere with the normal development of the others, thus leading to an undifferentiated dyscalculic pattern, even if the underlying neurological damage is limited. Secondly, the physical proximity of the areas involved makes it likely that there is a high correlation between impairment in one area and impairment in another. As pointed out by Dehaene et al. (2003) this explains why in the neuropsychological literature it is hard to find “pure” cases of verbal or number sense acalculia: the proximity of the parietal areas involved means that they are likely to be lesioned together. It is possible that this could occur also in developmental dyscalculia; for instance a gene-influenced growth factor could impact development in a whole subsection of the cortex, such as the inferior parietal lobule.

Normal infant and child numerical cognition

Of course our ability to make predictions about developmental dyscalculia from the adult neuropsychology and imaging literature rests on the assumption that numerical cognition systems in the adult and the child are similar. However, it is not yet clear whether this is the case, and we have little knowledge of how the infant system develops into the child and then the adult system (Ansari & Karmiloff-Smith, 2002). In this section we briefly review some of our key knowledge about the infant and child systems.

Number sense: our core magnitude representation

One obvious difference between the infant and adult numerical cognition system is that the infant is not born with an innate ability to process symbolic numerical codes, such as digits and number names. It was initially thought that all numerical knowledge had to be constructed through sensori-motor interaction with the environment (Piaget, 1952), but due to the many studies on infant numerical cognition in the past quarter of a century, we now know that infants are born with an ability to represent, discriminate, and operate on numerosities, although with only a limited degree of precision (For a recent review see Feigenson, Dehaene,

& Spelke, 2004). For instance, even with continuous visual cues such as luminance and occupied area controlled, 6-month old infants can discriminate between groups of 8 and 16 or 16 and 32 dots, but not 16 and 24 dots (Xu, Spelke, & Goddard, 2005; Xu & Spelke, 2000). Recent work by McCrink and Wynn (2004) has shown that 9-month-olds can approximately add and subtract collections of objects (for example 5 + 5 or 10 - 5). These approximate representations of number in infants are constrained by the ratio of the two numbers, and improve in precision during the first year of life (Lipton & Spelke, 2003).

Although as of yet no imaging evidence is available on the source of these representations in infants, due to the practical difficulties of conducting such studies, these characteristics are similar to the approximate representation present in animals (Gallistel & Gelman, 2000; Nieder, 2005), and in the HIPS of the adult human (see discussion above). Behavioral studies (and one ERP study) in pre-school children also support this conclusion (Berch, Foley, Hill, & Ryan, 1999; Girelli, Lucangeli, & Butterworth, 2000; Huntley-Fenner, 2001; Rubinsten, Henik, Berger, & Shahar-Shalev, 2002; Siegler & Opfer, 2003; Temple & Posner, 1998; see Noël, Rousselle, & Mussolin, 2005 for a review). The fact that this "number sense" system matures in the first year of life and is a core aspect of adult numerical cognition makes it a likely candidate for a core deficit in dyscalculia.

A second non-symbolic core system: object files

For some time, opponents of the concept of an innate numerosity representation have argued that infants were able to represent number based on their visuo-spatial abilities (e.g. Simon, 1999). It has now become clear that to keep track of small quantities of objects, infants are also able to use a visuo-spatial "object file" based system, which allows them to keep "pointers" to up to 3 or 4 objects, and also their continuous properties. Infants in fact may sometimes show a preference for using this system over the approximate magnitude system (Feigenson, 2005; Feigenson, Carey, & Hauser, 2002; Feigenson, Carey, & Spelke, 2002; Xu, 2003). It has been argued that adults may still use this system for subitizing, or rapidly identifying 3-4 objects (Trick & Pylyshyn, 1994), although this has not yet been proven to be the case (Feigenson et al., 2004). It is thus not certain whether and to what extent impairments in this second system could be responsible for developmental dyscalculia.

Development of symbolic capabilities

In addition to these two core systems children also develop (in the appropriate cultural context) the ability to represent numbers in a symbolic fashion, first using words, and later using arabic digits. This then allows them to extend their approximate abilities into the realm of exact arithmetic. Little developmental data is available on the neural foundations of developing symbolic representations, particularly on how these might be linked with the non-symbolic core systems discussed above. However it is clear that an impairment in symbolic representation or an impairment in the link between the non-symbolic and the symbolic are both possible causes for dyscalculia.

The first key development which occurs between the ages of 2 to 4 years in normal children is the acquisition of counting. In order to make full use of the counting procedure children need to learn the sequence of count words, and to understand and execute their one-to-one mapping to a set of objects. Finally they need to understand that this procedure gives the cardinality of the set. Some argue that it is only the language and procedural execution aspects of the task which need to be acquired, and that the concepts of one-to-one correspondence and cardinality comprehension are already present (Fuson, 1988; Wynn, 1990), whereas others claim that all of these concepts need to be acquired (Cordes & Gelman, 2005; Gelman & Gallistel, 1978). Wynn (1990), in particular, demonstrates that between 2 ½ and 3 1/2 years old , children exhibit a radical change in their understanding of counting and in their ability to use counting in simple quantity tasks.

Early work by Siegler and colleagues showed that prior to entering school children spontaneously count on their fingers, and can use them for simple additions (Siegler & Jenkins, 1989; Siegler & Shrager, 1984). Pre-school children have also usually mastered a strategy for exact addition, "counting all", in which for example, to add 2+4 they would count out two on their fingers, count out four using other fingers, and finally count all the fingers to give six. Eventually this strategy is replaced by a more advanced one, "counting on", in which children start at the first number, and count up the second number of units, e.g. "two...three, four, five six". Finally children learn the "min" strategy of starting at the largest number so that they have to count only the smallest distance, e.g. "four... five, six". As children get older, counting (with or without fingers) comes to play a decreasing role in addition, as memory-based retrieval strategies take over. However, it has been shown that even adults still rely on backup strategies in the event of a retrieval failure, such as decomposition (e.g. $8 + 6 \rightarrow (8 + 2) + 4 = 10 + 4 = 14$), or derivation from related facts (e.g. $8 + 6 \rightarrow (9 + 6) - 1 = 15 - 1 = 14$). (For a review see LeFevre, Smith-Chant, Hiscock, Daley, & Morris, 2003)

Another possible candidate for dyscalculia is therefore an impairment in the symbolic system. Although as mentioned earlier there is little developmental data on the neural bases of this system, in the adult we know that at least part of it, especially that governing counting and retrieval of arithmetic facts, is verbal in nature (Spelke & Tsivkin, 2001). Neuroimaging evidence suggests that it is at least partially governed by perisylvian language areas (Stanescu-Cosson *et al.*, 2000). In the case of strategy execution, there is currently no imaging data available even in the adult, however we can tentatively say that this ability is likely to be linked to the frontal lobes. This then generates two hypotheses for dyscalculia: a subtype linked to verbal impairment, and a subtype linked to executive dysfunction.

A further important hypothesis should be mentioned here. From the previous discussion it is evident that there are many bases for the representation of number, which can be divided into two broad categories, non-symbolic and symbolic. These representations clearly come from different sources; children are born with an approximate non-symbolic representation of number which is similar to that present in animals, and then they later learn an exact symbolic representation. Another obvious hypothesis for dyscalculia is that of a disconnection syndrome: dyscalculic children could have an intact non-symbolic representation of quantity, but fail to make the link between this and their newly acquired symbolic representations.

Summary of hypotheses for developmental dyscalculia

In summary of the previous sections, we now list the hypotheses put forward for developmental dyscalculia based on the neuropsychology, adult numerical cognition and developmental numerical cognition literature. There are two sets of questions to be addressed: firstly, whether there is a core deficit or not, and if so what type of dyscalculia would be associated with it, and secondly, what other subtypes of dyscalculia might be found.

We propose that if there is a single "core deficit" which causes dyscalculia, it is likely to be due to one of the following:

1. A deficit in number sense, or non-symbolic representation of number. This deficit would be caused by structural or functional impairment to the HIPS region of the intra-parietal sulcus. Its symptoms would include impaired understanding of the meaning of numbers, deficits in tasks which involve this area (non-symbolic tasks such as comparison of and approximate addition of dots, but also the symbolic tasks of numerical comparison, addition and subtraction), and reduced automatic activation of quantity from number words and digits. Because these are such basic deficits, they would be likely to cause a developmental delay in all aspects of math, except the highly verbal processes of counting and fact retrieval.
2. Impaired connections between symbolic and non-symbolic representations. In this case, we would expect to see the same pattern described above, but with one important

difference: little impairment on non-symbolic tasks.

Based on our literature review, we might also expect several other subtypes of developmental dyscalculia:

1. A deficit in verbal symbolic representation, related to impairment to the angular gyrus, the left inferior frontal and/or temporal language areas, or the left basal ganglia. This would result in difficulties learning and retrieving arithmetical facts (particularly for multiplication), and possibly also in learning the counting sequence.

2. A deficit in executive dysfunction, due to frontal dysfunction. This would also be likely to result in difficulties in arithmetical fact retrieval, but would furthermore result in difficulties in strategy and procedure usage.

3. A deficit in spatial attention, due to posterior superior parietal dysfunction. This type of deficit could be linked to the "object file" tracking system, and might therefore result in difficulties in subitizing (if this is the system underlying subitizing). It might also result in difficulties in perception of non-symbolic quantity information, and in quantity manipulation. However, due to the close intertwining of spatial and numerical representations, this subtype might be difficult to separate from a number sense subtype.

Developmental dyscalculia

Bearing in mind these hypotheses, we now turn to a review of the developmental dyscalculia literature, and examine to what extent we find them supported. Firstly we briefly discuss two important issues which should be borne in mind when thinking about this research, and which can make it difficult to compare results across different studies.

Identification

In the introductory section, we discussed the definition of developmental dyscalculia, a disorder in mathematical abilities presumed to be due to a specific impairment in brain function. In the educational field it has been traditional to identify learning disabilities by using standardized educational tests and defining dyscalculia as a significant lag in performance, taking into account age and IQ (e.g. Geary, Hamson, & Hoard, 2000; Jordan, Hanich, & Kaplan, 2003). Alternatively, some research laboratories studying dyscalculia have used their own tests based on neuropsychological batteries (e.g. Gross-Tsur et al., 1996). One important issue is the cutoff used to identify children as dyscalculic, which is essentially arbitrary, as in the case of dyslexia. This has lead to important differences in the populations of children studied (See Butterworth, 2005 for a detailed discussion), making it difficult to be certain of the true symptoms of dyscalculia.

Comorbid disorders

Two important disorders which appear to be comorbid with dyscalculia are dyslexia and attention deficit hyperactivity disorder (ADHD). Estimations for the comorbidity rate of dyslexia vary wildly, possibly due to differences in criteria, methodology and school year. For instance a longitudinal prevalence study by Badian (1999) found that 60% of persistent low arithmetic achievers were also low reading achievers (using a cut-off of the 25th percentile on average achievement over a 7-8 year period). Lewis et al. (1994) found a comorbidity rate (64%) of a similar order in their prevalence study, in which they used a cut-off of the 16th percentile for reading and math difficulties. At the other extreme, Gross-Tsur et al. (1996) found a dyslexia-dyscalculia comorbidity of only 17% in their sample of dyscalculics. However, their cut-off for dyslexia was the 5th percentile on a standardized reading and spelling test, which was much more conservative than that of the previous authors. The comorbidity rate of dyscalculia and ADHD is no less certain, having been addressed in only one large prevalence study (Gross-Tsur et al., 1996). These authors found that 26% of their dyscalculic sample showed symptoms of ADHD as measured by Connor's questionnaire.

The presence of comorbid disorders in dyscalculia is important to keep in mind, because studies have not always controlled for or reported them, and they may in fact be related to the symptoms observed.

Characteristics

Much work on the characteristics of dyscalculia has already been conducted in the educational field, although as we will discuss later, this work can be difficult to compare to the numerical cognition field. Here we review key findings in the educational field briefly; for a more in-depth discussion, the reader is referred to excellent reviews by Geary (1993, 2004). Several key characteristics of dyscalculia have been extensively studied, and are generally agreed on. Firstly, Geary and colleagues (e.g. Geary, Bow-Thomas, & Yao, 1992; Geary et al., 2000; Geary, Hoard, & Hamson, 1999) have consistently found an early delay in understanding some aspects of counting (order irrelevance, detection of double counts) amongst first and second grade children with dyscalculia. It is unknown whether these deficits, or other deficits in counting, continue after this age. Secondly and probably relatedly, many studies have reported a developmental delay in using counting strategies in simple addition, for instance dyscalculic children persist in using "counting all" strategies whilst their peers have learnt to "count on" (e.g. Geary, 1990; Geary, Brown, & Samaranayake, 1991; Geary et al., 2000; Jordan & Montani, 1997). Finally, a delay and persistent deficit in acquiring and using verbal facts has been well documented; dyscalculic children tend to keep using time-absorbing finger counting strategies for simple arithmetic facts that their peers have long since memorized (e.g. Ginsburg, 1997; Jordan & Montani, 1997; Kirby & Becker, 1988). A series of excellent longitudinal studies by Ostad (1997, 1999) suggests that this difference persists up until at least 5^{th} grade for addition and 7^{th} grade for subtraction.

A more controversial general deficit which has been proposed is a deficit in various components of working memory (Geary, 2004; Koontz & Berch, 1996; McLean & Hitch, 1999; Temple & Sherwood, 2002). Several studies conducted have found impairment on central executive tasks (D'Amico & Guarnera, 2005; Gathercole & Pickering, 2000; McLean

& Hitch, 1999; Passolunghi & Siegel, 2004), suggesting a possible frontal dysfunction, which fits with the procedural deficits discussed above. However, only some studies have found verbal working memory impairments (Wilson & Swanson, 2001), whereas others have not (McLean & Hitch, 1999; Passolunghi & Siegel, 2004). This may be because of different measures used for verbal working memory, particularly whether a digit span task is used. A recent study by D'Amico and Guarnera (2005) examined children with a thorough battery of tests for all three types of working memory, and found that dyscalculic children showed a deficit in digit span, but not pseudo-word span, suggesting that the deficit is in the representation of numerical information, rather than the representation or rehearsal of verbal information in general. Most earlier studies did not examine spatial working memory, however some more recent studies have found deficits in this domain too (D'Amico & Guarnera, 2005; Gathercole & Pickering, 2000; McLean & Hitch, 1999).

In our view, it is unlikely that working memory deficit(s) in themselves are the core deficit(s) in dyscalculia, but rather that both are co-occurring symptoms of other numerical, verbal, or spatial impairments[ii]. For example an impairment in the ability to store numerical information could result in dyscalculia, a reduced digit span, and possibly a lower central executive score (but solely for tasks involving numerical information). An impairment in the ability to shift spatial attention might result in dyscalculia, a reduced spatial span, and possibly a lower central executive score (for tasks involving spatial information). As we have seen, predictions such as these are starting to be examined, although it may be some time before the issue is clarified.

Dyscalculia subtypes

It is important to note that not all dyscalculic children show difficulties in all of the areas mentioned above, and that many authors have made the case for specific subtypes of developmental dyscalculia. Indeed the existence of known subtypes could lead to a clarification of symptoms: the current long list of characteristics might in fact be a mixture of many different subtypes. In this section we briefly review major subtype proposals which have been subject to or are based on several research studies.

Rourke and colleagues conducted much of the early subtyping research (see Rourke, 1993; Rourke & Conway, 1997 for reviews), and argued for two subtypes of mathematical disabilities: a verbal type, associated with left hemisphere impairment, and a spatial type, associated with right hemisphere impairment. They grouped dyscalculic children based on whether they had concurrent reading and spelling deficits (RDSD), or isolated arithmetic deficits (MD). Neuropsychological tests on these groups revealed a double dissociation: RDSD children performed better on visuo-spatial tests, and worse on verbal tests, whereas MD children showed the opposite pattern (Rourke & Finlayson, 1978). Further studies found that MD children also showed deficits in psychomotor and tactile-perceptual tasks, and on complex non-verbal abstract reasoning tasks (Rourke & Strang, 1978). However not all independent studies have been able to find evidence for these two subtypes of dyscalculia, for example Share et al. (1988) found this pattern only in boys and not in girls, and Shalev et al. (1997) failed to find it at all.

More recent studies by Jordan and colleagues (Jordan & Hanich, 2000; Jordan et al., 2003; Jordan & Montani, 1997) have also focused on grouping children into those with mathematical and reading disabilities (MDRD) and those with only mathematical disabilities (MD). However, unlike in the previous studies, these authors then measured performance on basic numerical and mathematical tasks. In general the results reveal a single dissociation: MDRD children are consistently worse than MD children in exact calculation, and solving story problems. However, there are no tasks at which MD children are worse than MDRD children, leaving open the possibility that the differences are simply due to the difficulty of the tasks. Furthermore, as of yet, other researchers using core numerical cognition tasks have failed to find a difference between the performance of MDRD and MD children (e.g. Landerl, Bevan, & Butterworth, 2004).

A third influential proposal, based on a synthesis of the educational and neuropsychological literature, has been made by Geary (1993; Geary, 2004), who posits three key subtypes of mathematical disabilities. The first is a procedural subtype, in which children show a delay in acquiring simple arithmetic strategies, and which Geary proposes may be a result of verbal working memory deficits, but perhaps also deficits in conceptual knowledge. The second is a semantic memory subtype, in which children show deficits in retrieval of facts, and which Geary proposes is due to a long term memory deficit. As discussed earlier, there is much evidence for procedural and fact retrieval deficits in dyscalculic children. The third and final subtype proposed by Geary is a visuo-spatial subtype, in which children show deficits in the spatial representation of number. However, there is little evidence for the existence of this subtype, although this may be due to the infrequency of testing for spatial abilities.

Behavioral and neural evidence for a number sense deficit in dyscalculia

From this overview of the dyscalculia literature, we turn now to a more in-depth evaluation of the evidence for core cognitive deficits in dyscalculia, particularly that of number sense. However, first we note that it is difficult to use research in special education for this purpose. This is because, as pointed out by other authors (e.g. Ansari & Karmiloff-Smith, 2002), many educational studies have not used basic measures of numerical cognition, but rather higher level tests. These tests are likely to involve many combinations of cognitive processes, and thus may not reveal specific numerical deficits. Where low-level tasks have been used, the authors have not always used reaction time measures, which may reveal abnormalities where accuracy does not (Butterworth, 2005; Jordan & Montani, 1997). Thus below we discuss mostly relevant research from clinical neuropsychology.

An important exception to this rule is a recent study by Llanderl et al. (2004), whose results provide preliminary support for the “number sense” core deficit hypothesis. The authors tested a group of 21 eight and nine year old dyscalculic children and compared their performance on core number processing tasks to that of 18 controls. They found that the dyscalculic group showed a deficit in speed of number comparison, although their performance on a non numerical comparison task was normal. They also found that dyscalculic children showed a steeper increase in reaction time than controls when enumerating small quantities of dots, suggesting an impairment in subitizing, as had been suggested by earlier data (Koontz & Berch, 1996). In the counting range a similar steeper increase in reaction time was found[iii].

In the context of clinical neuropsychology, there are only a few published developmental dyscalculia cases which support the core deficit hypothesis. For instance, Butterworth (1999) reports the case of “Charles”, a dyscalculic adult who despite normal IQ and reasoning shows a deficit in numerical comparison (even with a reverse distance effect) and in subitizing. Kaufmann (2002) reports a similar case of a 14-year-old boy who showed no distance effect (although he did show normal subitizing), even though he was perfectly able to complete multidigit calculation procedures. Both of these cases also showed a large amount of finger counting as opposed to using mental strategies and retrieval.

Other cases of developmental dyscalculia have been reported in the context of a “developmental Gerstmann’s syndrome” (Benson & Geschwind, 1970; Kinsbourne & Warrington, 1963), in which children show left-right disorientation, finger agnosia, agraphia and dyscalculia, in the context of normal intelligence, although not always normal reading performance. Children with this syndrome show little difficulty with fact retrieval, but difficulties in addition, and particularly subtraction, which is consistent with a number sense deficit. However, as in the adult research, the existence of the syndrome as a coherent whole as been questioned (Miller & Hynd, 2004; Spellacy & Peter, 1978).

It should be noted that several case studies do support the possible existence of different subtypes of developmental dyscalculia. For instance, Kaufmann's (2002) case appears to have more than just a quantity representation deficit, because he also showed a large impairment in retrieval of arithmetical facts which seemed to be due to long term memory interference, possibly caused by executive dysfunction. Three cases published by C. M. Temple (1989, 1991) support the existence of other subtypes of dyscalculia, seemingly independent of number sense: a procedural deficit related to frontal damage, a fact retrieval deficit in the presence of phonological dyslexia, and a transcoding deficit in the presence of impaired verbal working memory.

Finally we turn to neural evidence of deficits in dyscalculia, which although in its infancy, is promising for the hypothesis of a number sense deficit. Several recent studies in specific subpopulations of dyscalculics have implicated abnormalities in the intra-parietal sulcus as would be predicted. Isaacs et al. (2001) selected two groups of 12 adolescents who had been born preterm (matched for IQ), one group showing impairment in arithmetic, and the other not. The authors compared the density of gray matter between the two groups of adolescents, and found that only the left IPS showed reduced grey matter in the arithmetically-impaired group, at the precise coordinates of the horizontal intra-parietal sulcus. (See Figure 2a).

Likewise, Molko and colleagues (Bruandet, Molko, Cohen, & Dehaene, 2004; Molko et al., 2003) studied women with Turner's syndrome (X monosomy), for which mathematical learning difficulties have been consistently reported (e.g. Mazzocco & McCloskey, 2005; e.g. Rovet, Szekely, & Hockenberry, 1994; Temple & Marriott, 1998). Bruandet et al. used a testing battery of symbolic and non-symbolic tasks, and found that Turners subjects showed deficits in number sense tasks, such as cognitive estimation, subitizing, addition and subtraction. In a further imaging study, Molko et al. observed a disorganization of the right IPS, which was of abnormal depth. Furthermore, fMRI revealed reduced activation in the right IPS as a function of number size during exact calculation. (See Figure 2b).

*** INSERT FIGURE 2 AROUND HERE ***

A similar reduction in normal activation levels, extending to a broader parietoprefrontal network, has been observed in at least one other genetic condition associated with dyscalculia, fragile X (Rivera, Menon, White, Glaser, & Reiss, 2002). Other genetic conditions such as velocardiofacial syndrome, may show similar impairments (Eliez et al., 2001; Simon et al., 2002; Simon et al., 2005).

Conclusions

Although much research remains to be done, the preliminary evidence supporting the role of a number sense deficit in dyscalculia is promising. Behavioral evidence suggests that dyscalculics show impairments in numerical comparison and subitizing, which would be expected from an impairment in number sense. Neural evidence, although as of yet only from special populations, points to the role of the HIPS, which is believed to represent quantity. Furthermore dyscalculic children have been reported as showing persistent difficulties in learning simple addition and subtraction strategies, which would fit with a reduced understanding of the meaning of numbers, or ability to manipulate them.

However whether an impairment of number sense is a core deficit responsible for dyscalculia is not clear. Nor are we currently able to distinguish between a deficit in number sense itself or in its connections to symbolic representations of numerosity. The possibility of multiple types of dyscalculia remains an important one, with supporting information from special education research and from clinical case studies. In particular, there is much evidence for fact retrieval deficits, which have been observed in isolation in case studies and which are consistently observed in dyscalculic populations. This deficit could be due to either a verbal memory deficit or a deficit in executive function. In the case of the former, the comorbidity between dyslexia and dyscalculia may be an important factor, whereas for the latter, that between ADHD and dyscalculia may be important. The possibility of a spatial attention deficit subtype of dyscalculia remains an important one, which should be investigated.

These possibilities should be borne in mind by researchers in the dyscalculia and mathematical disabilities fields. We suggest that future studies in these areas test children for dyslexia, ADHD, executive function, and spatial attention, in order to allow an analysis of results by possible subtypes.

Implications for education and intervention

In this final section, we discuss recommendations for the field of education. We highlight the importance of the cognitive neuroscience and educational fields working more closely together in order to try and achieve three key aims: (1) the development of a "neurocognitive" description of dyscalculia, (2) the development, norming and educational use of core tests based on numerical cognition research, and (3) the development and testing of new educational remediation methods.

Towards a "neurocognitive" description of dyscalculia

By a neurocognitive description of dyscalculia, we mean a description which is based on, and ideally measured by, behavioral and neuroimaging paradigms previously studied in normal subjects, and with a theoretical basis in numerical cognition. Such a description would specify at the neural level the brain systems implicated in dyscalculia, and at the behavioral level specific cognitive deficits which would be expected as a result of neural impairments. We argue that this would allow for better identification, better treatment, and the possibility of prevention. Better identification might one day mean being able scan children for brain function, and immediately identify the subtype of dyscalculia present. Better treatment could mean designing a custom-built targeted remediation which would give each child the greatest chance of success. The possibility of prevention depends on early identification, and on the plasticity of the brain circuits involved, but early identification may be possible for dyslexia (Lyytinen et al., in press), and might also be for dyscalculia.

Core Ability Tests

To the extent that dyscalculia is caused by or correlated with deficits in core numerical cognition processes, it is important to test for it with batteries of basic numerical cognition tasks. These batteries should measure reaction time as well as accuracy, and include symbolic and well as non-symbolic tasks. The presence of personal computers in most western classrooms now makes this feasible, and indeed, some countries are already starting to move to this system for testing for dyscalculia, for instance in the United Kingdom, Brian Butterworth's "Dyscalculia Screener" (Butterworth, 2003) is now being used by some schools to identify dyscalculic children on the basis of dot enumeration and number comparison. Which tests should be used in such instruments is still an issue which is under discussion, and should be informed by ongoing research. Thus far, subitizing and number comparison are good candidates.

New Remediation Methods

Identifying neurocognitive deficits and subtypes of dyscalculia should allow for new remediation methods to be developed. This has already been the case in the field of dyslexia, as discussed in the introductory section. Based on what we know about dyscalculia, we hypothesize that remediation techniques based on number sense training should be effective. As of yet it is difficult to say whether techniques based on verbal memory training, visuo-spatial attention or executive attention training would also be effective, but this is a possibility.

*** INSERT FIGURE 3 AROUND HERE ***

In our laboratory, we have developed and tested an adaptive computer game remediation, similar in concept to those used in dyslexia. This software, “The Number Race”, is based on the number sense core deficit hypothesis, and is designed to provide intensive training on a key number sense task: numerical comparison, and to reinforce links between non-symbolic and symbolic representations of number. (See Figure 3). The software adapts to children’s performance by increasing the difficulty of the numerical comparison, by imposing a variable speed limit, and by increasing the ratio of symbolic to non-symbolic stimuli according to their performance. An early “open-trial” pilot study with this software shows promising results, with children showing significant improvements in subitizing, subtraction, and numerical comparison. Whether this tool will ultimately prove useful for all children with developmental dyscalculia is a matter for further research.

Overall Conclusions

Developmental dyscalculia is a disorder in mathematical abilities presumed to be due to impaired brain function. It appears to have a similar prevalence to its equivalent in reading (dyslexia), but is vastly understudied in comparison. Its basic behavioral symptoms and its neurological bases are only just starting to be investigated. In this chapter, we proposed possible causes of dyscalculia from reviewing the literature on the neurological bases of adult numerical cognition and on development of numerical cognition in children. We identified two possible causes of a “core deficit”: 1) a deficit in number sense, or non-symbolic representation of number, related to an impairment in the horizontal intra-parietal sulcus (HIPS) area, and 2) a failure to build adequate connections between non-symbolic and symbolic representations of number. We also identified three other possible causes of different subtypes of dyscalculia; deficits in verbal symbolic representation, executive dysfunction or spatial attention.

We then reviewed what is currently known about dyscalculia to examine which of these hypotheses are supported by current data. Research conducted in the education field has identified several key deficits in dyscalculic children's acquisition of counting, counting-based strategies, and verbal facts. Some researchers in this field have argued that dyscalculia can be divided into verbal and non-verbal subtypes, however research results have not always supported this proposal. One of the problems is that education research has typically used higher-level tasks composed of many component processes. We emphasize the need for future studies of dyscalculia symptoms to use a wide variety of tasks, including low-level numerical cognition tasks, and non-symbolic as well as symbolic tasks.

The evidence which is available using low-level numerical cognition tasks provides preliminary support for the "number sense" core deficit hypothesis. Dyscalculic children show impairments in numerical comparison and subitizing, and research in special populations suggests that this may be linked to an underfunctioning of the HIPS, which is known to represent quantity. However this research is in its infancy, and much more is needed for the issue to be resolved.

We finally discussed implications for education and intervention, and emphasized three key aims. The first is the development of a "neurocognitive" description of dyscalculia, which would allow for better identification, treatment and possibly prevention of dyscalculia. The second is the development of core ability tests which would be based on the neurocognitive description developed. The third is the development of new remediation methods, which target children's core deficits.

References

American Psychiatric Association. (1994). *Diagnostic and statistical manuel of mental disorders dsm-iv*:American Psychiatric Pub.
Ansari, D., & Karmiloff-Smith, A. (2002). Atypical trajectories of number development: A neuroconstructivist perspective. *Trends in Cognitive Sciences, 6*(12), 511-516.
Badian, N. A. (1983). Dyscalculia and nonverbal disorders of learning. In H. R. Myklebust (Ed.), *Progress in learning disabilities* (Vol. 5, pp. 235-264). New York: Stratton.
Badian, N. A. (1999). Persistent arithmetic, reading, or arithmetic and reading disability. *Annals of Dyslexia, 49*, 45-70.
Benson, D. F., & Geschwind, N. (1970). Developmental gerstmann syndrome. *Neurology, 20*(3), 293-298.
Benton, A. L. (1992). Gerstmann's syndrome. *Archives of Neurology, 49*(5), 445-447.
Berch, D. B., Foley, E. J., Hill, R. J., & Ryan, P. M. (1999). Extracting parity and magnitude from arabic numerals: Developmental changes in number processing and mental representation. *Journal of Experimental Child Psychology, 74*(4), 286.
Bruandet, M., Molko, N., Cohen, L., & Dehaene, S. (2004). A cognitive characterization of dyscalculia in turner syndrome. *Neuropsychologia, 42*(3), 288-298.
Butterworth, B. (1999). *The mathematical brain*.London: Macmillan.
Butterworth, B. (2003). Dyscalculia screener. London: nferNelson.
Butterworth, B. (2005). The development of arithmetical abilities. *Journal of Child Psychology and Psychiatry, 46*(1), 3-18.
Butterworth, B. (2005). Developmental dyscalculia. In J. Campbell (Ed.), *Handbook of mathematical cognition*.New York: Psychology Press.
Cappelletti, M., Butterworth, B., & Kopelman, M. (2001). Spared numerical abilities in a case of semantic dementia. *Neuropsychologia, 39*(11), 1224.
Chochon, F., Cohen, L., van de Moortele, P. F., & Dehaene, S. (1999). Differential contributions of the left and right inferior parietal lobules to number processing. *Journal of Cognitive Neuroscience, 11*(6), 617-630.
Cohen, L., Wilson, A. J., Izard, V., & Dehaene, S. (in press). Acalculia and gerstmann's syndrome. In O. Godefroy & J. Bogousslavsky (Eds.), *Cognitive and behavioral neurology of stroke*:Cambridge University Press.
Cordes, S., & Gelman, R. (2005). The young numerical mind. In J. D. Campbell (Ed.), *Handbook of mathematical cognition* (pp. 127-142). New York: Psychology Press.
D'Amico, A., & Guarnera, M. (2005). Exploring working memory in children with low arithmetical achievement. *Learning and Individual Differences, 15*(3), 189.
Dehaene, S. (1997). *The number sense: How the mind creates mathematics*.Oxford: Oxford University Press.
Dehaene, S. (2001). Précis of the number sense. *Mind and Language, 16*, 16-36.
Dehaene, S., & Cohen, L. (1997). Cerebral pathways for calculation: Double dissociation between rote verbal and quantitative knowledge of arithmetic. *Cortex, 33*(2), 219-250.
Dehaene, S., Piazza, M., Pinel, P., & Cohen, L. (2003). Three parietal circuits for number processing. *Cognitive Neuropsychology*(20), 487-506.
Dehaene, S., Spelke, E., Pinel, P., Stanescu, R., & Tsivkin, S. (1999). Sources of mathematical thinking: Behavioral and brain-imaging evidence. *Science, 284*(5416), 970-974.
Dehaene, S., Tzourio, N., Frak, V., Raynaud, L., Cohen, L., Mehler, J., et al. (1996). Cerebral activations during number multiplication and comparison: A pet study. *Neuropsychologia, 34*(11), 1097-1106.
Delazer, M., Karner, E., Zamarian, L., Donnemiller, E., & Benke, T. (in press). Number

processing in posterior cortical atrophy--a neuropsycholgical case study. *Neuropsychologia, In Press, Corrected Proof.*
Eden, G. F. (2002). The role of neuroscience in the remediation of students with dyslexia. *Nature Neuroscience, 5*(Supplement), 1080-1084.
Eger, E., Sterzer, P., Russ, M. O., Giraud, A.-L., & Kleinschmidt, A. (2003). A supramodal number representation in human intraparietal cortex. *Neuron, 37*(4), 719.
Eliez, S., Blasey, C. M., Menon, V., White, C. D., Schmitt, J. E., & Reiss, A. L. (2001). Functional brain imaging study of mathematical reasoning abilities in velocardiofacial syndrome (del22q11.2). *Genetics in Medicine, 3*(1), 49-55.
Feigenson, L. (2005). A double-dissociation in infants' representations of object arrays. *Cognition, 95*(3), B37.
Feigenson, L., Carey, S., & Hauser, M. (2002). The representations underlying infants' choice of more: Object files versus analog magnitudes. *Psychological Science, 13*(2), 150-156.
Feigenson, L., Carey, S., & Spelke, E. (2002). Infants' discrimination of number vs. Continuous extent. *Cognitive Psychology, 44*(1), 33-66.
Feigenson, L., Dehaene, S., & Spelke, E. (2004). Core systems of number. *Trends in Cognitive Sciences, 8*(7), 307-314.
Fuson, K. C. (1988). *Children's counting and concepts of number.*New York: Springer-Verlag.
Gallistel, C. R., & Gelman, R. (2000). Non-verbal numerical cognition: From reals to integers. *Trends in Cognitive Sciences, 4*(2), 59-65.
Gathercole, S. E., & Pickering, S. J. (2000). Working memory deficits in children with low achievements in the national curriculum at 7 years of age. *British Journal of Educational Psychology, 70*(2), 177-194.
Geary, D. C. (1990). A componential analysis of an early learning deficit in mathematics. *Journal of Experimental Child Psychology, 49*(3), 363-383.
Geary, D. C. (1993). Mathematical disabilities: Cognitive, neuropsychological and genetic components. *Psychological Bulletin, 114*(2), 345-362.
Geary, D. C. (2004). Mathematics and learning disabilities. *Journal of Learning Disabilities, 37*(1), 4-15.
Geary, D. C., Bow-Thomas, C. C., & Yao, Y. (1992). Counting knowledge and skill in cognitive addition: A comparison of normal and mathematically disabled children. *Journal of Experimental Child Psychology, 54*(3), 372-391.
Geary, D. C., Brown, S. C., & Samaranayake, V. A. (1991). Cognitive addition: A short longitudinal study of strategy choice and speed-of-processing differences in normal and mathematically disabled children. *Developmental Psychology, 27*(5), 787-797.
Geary, D. C., Hamson, C. O., & Hoard, M. K. (2000). Numerical and arithmetical cognition: A longitudinal study of process and concept deficits in children with learning disability. *Journal of Experimental Child Psychology, 77*(3), 236-263.
Geary, D. C., Hoard, M. K., & Hamson, C. O. (1999). Numerical and arithmetical cognition: Patterns of functions and deficits in children at risk for a mathematical disability. *Journal of Experimental Child Psychology, 74*(3), 213-239.
Gelman, R., & Gallistel, C. R. (1978). *The child's understanding of number.*Cambridge Mass.: Harvard University Press.
Gersten, R., & Chard, D. (1999). Number sense: Rethinking arithmetic instruction for students with mathematical disabilities. *The Journal of special education, 33*(1), 18 (11 pages).
Gerstmann, J. (1940). Syndrome of finger agnosia disorientation for right and left agraphia and acalculia. *Archives of Neurology and Psychiatry, 44*, 398-408.

Ginsburg, H. P. (1997). Mathematics learning disabilities: A view from developmental psychology. *Journal of Learning Disabilities, 30*(1), 20-33.
Girelli, L., Lucangeli, D., & Butterworth, B. (2000). The development of automaticy of accessing number magnitude. *Journal of Experimental Child Psychology, 76*, 104-122.
Goswami, U. (2003). Why theories about developmental dyslexia require developmental designs. *Trends in Cognitive Sciences, 7*(12), 534.
Gross-Tsur, V., Manor, O., & Shalev, R. S. (1996). Developmental dyscalculia: Prevalence and demographic features. *Dev Med Child Neurol, 38*(1), 25-33.
Huntley-Fenner, G. (2001). Children's understanding of number is similar to adults' and rats': Numerical estimation by 5-7-year-olds. *Cognition, 78*(3), B27-B40.
Isaacs, E. B., Edmonds, C. J., Lucas, A., & Gadian, D. G. (2001). Calculation difficulties in children of very low birthweight: A neural correlate. *Brain, 124*(9), 1701-1707.
Jackson, M., & Warrington, E. K. (1986). Arithmetic skills in patients with unilateral cerebral lesions. *Cortex, 22*(4), 611-620.
Jordan, N. C., & Hanich, L. B. (2000). Mathematical thinking in second-grade children with different forms of ld. *Journal of Learning Disabilities, 33*(6), 567-578.
Jordan, N. C., Hanich, L. B., & Kaplan, D. (2003). A longitudinal study of mathematical competencies in children with specific mathematics difficulties versus children with comorbid mathematics and reading difficulties. *Child Development, 74*(3), 834-850.
Jordan, N. C., & Montani, T. O. (1997). Cognitive arithmetic and problem solving: A comparison and children with specific and general mathematics difficulties. *Journal of Learning Disabilities, 30*(6), 624-634.
Kaufmann, L. (2002). More evidence for the role of the central executive in retrieving arithmetic facts - a case study of severe developmental dyscalculia. *Journal of Clinical & Experimental Neuropsychology, 24*(3), 302-310.
Kinsbourne, M., & Warrington, E. K. (1963). The developmental gerstmann syndrome. *Archives of Neurology, 8*, 490-501.
Kirby, J. R., & Becker, L. D. (1988). Cognitive components of learning problems in arithmetic. *Remedial and Special Education, 9*(5), 7-16.
Koontz, K. L., & Berch, D. B. (1996). Identifying simple numerical stimuli: Processing inefficiencies exhibited by arithmetic learning disabled children. *Mathematical Cognition, 2*(1), 1-23.
Kosc, L. (1974). Developmental dyscalculia. *Journal of Learning Disabilities, 7*(3), 164-177.
Landerl, K., Bevan, A., & Butterworth, B. (2004). Developmental dyscalculia and basic numerical capacities: A study of 8-9-year-old students. *Cognition, 93*(2), 99-125.
Lee, K. M. (2000). Cortical areas differentially involved in multiplication and subtraction: A functional magnetic resonance imaging study and correlation with a case of selective acalculia. *Annals of Neurology, 48*(4), 657-661.
LeFevre, J.-A., Smith-Chant, B. L., Hiscock, K., Daley, K. E., & Morris, J. (2003). Young adults' strategic choices in simple arithmetic: Implications for the development of mathematical representations. In A. J. Baroody & A. Dowker (Eds.), *The development of arithmetic concepts and skills: Constructing adaptive expertise. Studies in mathematical thinking and learning* (pp. 203-228).
Lemer, C., Dehaene, S., Spelke, E., & Cohen, L. (2003). Approximate quantities and exact number words: Dissociable systems. *Neuropsychologia, 41*(14), 1942-1958.
Lewis, C., Hitch, G. J., & Walker, P. (1994). The prevalence of specific arithmetic difficulties and specific reading difficulties in 9- to 10-year old boys and girls. *Journal of Child Psychology & Psychiatry & Allied Disciplines, 35*(2), 283-292.
Lipton, J., & Spelke, E. (2003). Origins of number sense: Large-number discrimination in human infants. *Psychological Science, 14*(5), 396-401.

Lyytinen, H., Guttorm, T. K., Huttunen, T., Hamalainen, J., Leppanen, P. H. T., & Vesterinen, M. (in press). Psychophysiology of developmental dyslexia: A review of findings including studies of children at risk for dyslexia. *Journal of Neurolinguistics, In Press, Corrected Proof.*

Mazzocco, M. M. M., & McCloskey, M. (2005). Math performance in girls with turner or fragile x syndrome. In J. D. Campbell (Ed.), *Handbook of mathmatical cognition*.New York: Psychology Press.

McCrink, K., & Wynn, K. (2004). Large-number addition and subtraction by 9-month-old infants. *Psychological Science, 15*(11), 776-781.

McLean, J. F., & Hitch, G. J. (1999). Working memory impairments in children with specific arithmetic learning difficulties. *Journal of Experimental Child Psychology, 74*(3), 240-260.

Miller, C. J., & Hynd, G. W. (2004). What ever happened to developmental gerstmann's syndrome? Links to other pediatric, genetic, and neurodevelopmental syndromes. *J Child Neurol, 19*(4), 282-289.

Molko, N., Cachia, A., Riviere, D., Mangin, J. F., Bruandet, M., Le Bihan, D., et al. (2003). Functional and structural alterations of the intraparietal sulcus in a developmental dyscalculia of genetic origin. *Neuron, 40*(4), 847-858.

Nieder, A. (2005). Counting on neurons: The neurobiology of numerical competence. *Nature Reviews Neuroscience, 6*(3), 177.

Nieder, A., & Miller, E. K. (2004). A parieto-frontal network for visual numerical information in the monkey. *PNAS, 101*(19), 7457-7462.

Noël, M.-P., Rousselle, L., & Mussolin, C. (2005). Magnitude representation in children: Its development and dysfunction. In J. Campbell (Ed.), *Handbook of mathematical cognition*.New York: Psychology Press.

Ostad, S. A. (1997). Developmental differences in addition strategies: A comparison of mathematically disabled and mathematically normal children. *British Journal of Educational Psychology, 67*, 345-357.

Ostad, S. A. (1999). Developmental progression of subtraction strategies: A comparison of mathematically normal and mathematically disabled children. *European Journal of Special Needs Education, 14*(1), 21-36.

Passolunghi, M. C., & Siegel, L. S. (2004). Working memory and access to numerical information in children with disability in mathematics. *Journal of Experimental Child Psychology, 88*(4), 348.

Pesenti, M., Thioux, M., Seron, X., & De Volder, A. (2000). Neuroanatomical substrates of arabic number processing, numerical comparison, and simple addition: A pet study. *Journal of Cognitive Neuroscience, 12*(3), 461-479.

Piaget, J. (1952). *The child's conception of number*.New York: Norton.

Piazza, M., Izard, V., Pinel, P., Le Bihan, D., & Dehaene, S. (2004). Tuning curves for approximate numerosity in the human intraparietal sulcus. *Neuron, 44*, 547-555.

Pinel, P., Dehaene, S., Riviere, D., & LeBihan, D. (2001). Modulation of parietal activation by semantic distance in a number comparison task. *NeuroImage, 14*(5), 1013.

Pinel, P., Piazza, M., Le Bihan, D., & Dehaene, S. (2004). Distributed and overlapping cerebral representations of number, size, and luminance during comparative judgments. *Neuron, 41*(6), 983.

Rivera-Batiz, F. L. (1992). Quantitative literacy and the likelihood of employment among young adults in the united states. *The Journal of human resources, 27*(2), 313-328.

Rivera, S. M., Menon, V., White, C. D., Glaser, B., & Reiss, A. L. (2002). Functional brain activation during arithmetic processing in females with fragile x syndrome is related to fmr1 protein expression. *Human Brain Mapping, 16*, 206-218.

Robinson, C. S., Menchetti, B. M., & Torgesen, J. K. (2002). Toward a two-factor theory of one type of mathematics disabilities. *Learning Disabilities Research & Practice, 17*, 81.

Rosselli, M., & Ardila, A. (1989). Calculation deficits in patients with right and left hemisphere damage. *Neuropsychologia, 27*(5), 607-617.

Rourke, B. P. (1993). Arithmetic disabilities, specific and otherwise: A neuropsychological perspective. *Journal of Learning Disabilities, 26*(4), 214-226.

Rourke, B. P., & Conway, J. A. (1997). Disabilities of arithmetic and mathematical reasoning: Perspectives from neurology and neuropsychology. *Journal of Learning Disabilities, 30*(1), 34-46.

Rourke, B. P., & Finlayson, M. A. (1978). Neuropsychological significance of variations in patterns of academic performance: Verbal and visual-spatial abilities. *Journal of Abnormal Child Psychology, 6*(1), 121-133.

Rourke, B. P., & Strang, J. D. (1978). Neuropsychological significance of variations in patterns of academic performance: Motor, psychomotor, and tactile-perceptual abilities. *Journal of Pediatric Psychology, 3*, 62-66.

Rovet, J., Szekely, C., & Hockenberry, M.-N. (1994). Specific arithmetic calculation deficits in children with turner syndrome. *Journal of Clinical & Experimental Neuropsychology, 16*(6), 820-839.

Rubinsten, O., Henik, A., Berger, A., & Shahar-Shalev, S. (2002). The development of internal representations of magnitude and their association with arabic numerals. *Journal of Experimental Child Psychology, 81*(1), 74-92.

Shalev, R. S., & Gross-Tsur, V. (1993). Developmental dyscalculia and medical assessment. *Journal of Learning Disabilities, 26*(2), 134-137.

Shalev, R. S., & Gross-Tsur, V. (2001). Developmental dyscalculia. *Pediatric Neurology, 24*(5), 337-342.

Shalev, R. S., Manor, O., & Gross-Tsur, V. (1997). Neuropsychological aspects of developmental dyscalculia. *Mathematical Cognition, 3*(2), 105-120.

Share, D. L., Moffitt, T. E., & Silva, P. A. (1988). Factors associated with arithmetic-and-reading disability and specific arithmetic disability. *Journal of Learning Disabilities, 21*(5), 313-320.

Siegler, R. S., & Jenkins, E. A. (1989). *How children discover new strategies.*Hillsdale N.J.: Lawrence Erlbaum Associates.

Siegler, R. S., & Opfer, J. E. (2003). The development of numerical estimation: Evidence for multiple representations of numerical quantity. *Psychological Science, 14*(3), 237-243.

Siegler, R. S., & Shrager, J. (1984). Strategy choices in addition and subtraction: How do children know what to do? In C. Sophian (Ed.), *Origins of cognitive skill* (pp. 241-312). Hillsdale, NJ.: Erlbaum.

Simon, T. J. (1999). The foundations of numerical thinking in a brain without numbers. *Trends in Cognitive Sciences, 3*(10), 363.

Simon, T. J., Bearden, C. E., Moss, E. M., McDonald-McGinn, D., Zackai, E., & Wang, P. P. (2002). Cognitive development in vcfs. *Progress in Pediatric Cardiology, 15*(2), 109.

Simon, T. J., Ding, L., Bish, J. P., McDonald-McGinn, D. M., Zackai, E. H., & Gee, J. (2005). Volumetric, connective, and morphologic changes in the brains of children with chromosome 22q11.2 deletion syndrome: An integrative study. *NeuroImage, 25*(1), 169.

Spelke, E. S., & Tsivkin, S. (2001). Language and number: A bilingual training study. *Cognition, 78*(1), 45-88.

Spellacy, F., & Peter, B. (1978). Dyscalculia and elements of the developmental gerstmann syndrome in school children. *Cortex, 14*(2), 197-206.

Stanescu-Cosson, R., Pinel, P., Moortele, P.-F. v. d., Le Bihan, D., Cohen, L., & Dehaene, S. (2000). Understanding dissociations in dyscalculia: A brain imaging study of the impact of number size on the cerebral networks for exact and approximate calculation. *Brain, 123*(11), 2240-2255.

Tallal, P., Miller, S. L., Bedi, G., Byma, G., Wang, X., Nagarajan, S. S., et al. (1996). Language comprehension in language-learning impaired children improved with acoustically modified speech. *Science, 271*(5245), 81-84.

Temple, C. M. (1989). Digit dyslexia: A category-specific disorder in development dyscalculia. *Cognitive Neuropsychology, 6*(1), 93-116.

Temple, C. M. (1991). Procedural dyscalculia and number fact dyscalculia: Double dissociation in developmental dyscalculia. *Cognitive Neuropsychology, 8*(2), 155-176.

Temple, C. M., & Marriott, A. J. (1998). Arithmetical ability and disability in turner's syndrome: A cognitive neuropsychological analysis. *Developmental Neuropsychology, 14*(1), 47-67.

Temple, C. M., & Sherwood, S. (2002). Representation and retrieval of arithmetical facts: Developmental difficulties. *Quarterly Journal of Experimental Psychology A, 55A*(3), 733-752.

Temple, E., Deutsch, G. K., Poldrack, R. A., Miller, S. L., Tallal, P., Merzenich, M. M., et al. (2003). Neural deficits in children with dyslexia ameliorated by behavioral remediation: Evidence from functional mri. *Proc Natl Acad Sci U S A, 100*(5), 2860-2865.

Temple, E., & Posner, M. I. (1998). Brain mechanisms of quantity are similar in 5-year-old children and adults. *Proceedings of the National Academy of Science, 95*, 7836-7841.

Trick, L. M., & Pylyshyn, Z. W. (1994). Why are small and large numbers enumerated differently? A limited-capacity preattentive stage in vision. *Psychological Review, 101*(1), 80-102.

Wilson, K. M., & Swanson, H. L. (2001). Are mathematics disabilities due to a domain-general or a domain-specific working memory deficit? *Journal of Learning Disabilities, 34*(3), 237-248.

Wynn, K. (1990). Children's understanding of counting. *Cognition, 36*, 155-193.

Xu, F. (2003). Numerosity discrimination in infants: Evidence for two systems of representations. *Cognition, 89*(1), B15-B25.

Xu, F., Spelke, E., & Goddard, S. (2005). Number sense in human infants. *Developmental Science, 8*(1), 88-101.

Xu, F., & Spelke, E. S. (2000). Large number discrimination in 6-month-old infants. *Cognition, 74*(1), B1-B11.

Zago, L., Pesenti, M., Mellet, E., Crivello, F., Mazoyer, B., & Tzourio-Mazoyer, N. (2001). Neural correlates of simple and complex mental calculation. *Neuroimage, 13*(2), 314-327.

Footnotes

[i] See Geary and Hoard for a discussion of the similarities between the dyscalculia and mathematical learning disabilities literature.
[ii] Alternatively, an association could be purely circumstantial, due to the proximity of brain areas involved in working memory and numerical representation.
[iii] These results must be taken with caution, as the first was not significant, and the second only marginal, possibly due to a lack of statistical power.

Author Note

Corresponding author: Anna J. Wilson, INSERM-CEA Unit 562 « Cognitive Neuroimaging », Service Hospitalier Frédéric Joliot, CEA-DRM-DSV, Orsay, France. Email: ajwilsonkiwi@yahoo.fr. We kindly acknowledge financial support from the Fyssen Foundation (AJW), INSERM, and a McDonnell Foundation centennial fellowship (SD).

Figure 1

Three-dimensional representation of parietal regions activated in numerical tasks (see text for details). For better visualization, the clusters show all parietal voxels activated in at least 40% of studies in a given group (redrawn from Dehaene et al., 2003).

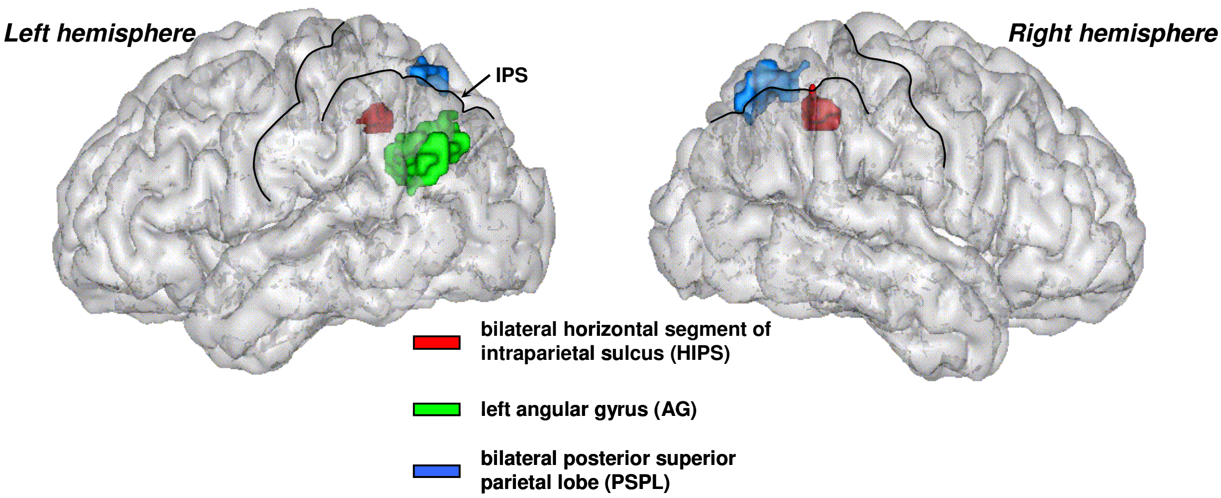

Figure 2

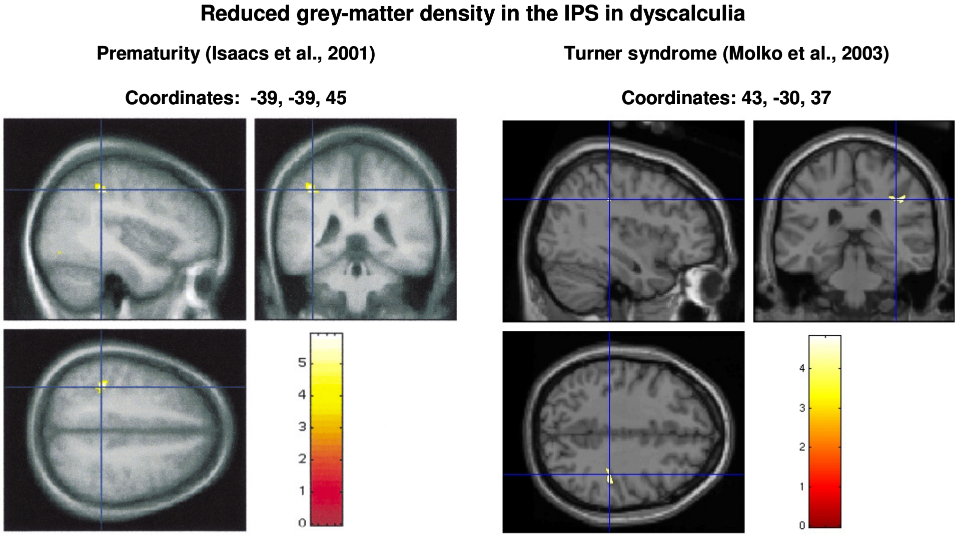
Reduced grey-matter density in the IPS in dyscalculia
Prematurity (Isaacs et al., 2001)
Coordinates: -39, -39, 45
Turner syndrome (Molko et al., 2003)
Coordinates: 43, -30, 37

Figure 3

Screenshots from « The Number Race », remediation software for dyscalculia in the form of an adaptive game (produced in our laboratory by Anna Wilson). The child plays the character of the dolphin, and has to choose the larger of two numerosities, before her competitor (the crab) arrives at the key and steals this many piece of gold. Here we see a high difficulty level with addition and subtraction required before numerical comparison is performed. The child then wins the same amount of squares on a game board, where she must avoid landing on anemone hazards. Once she arrives at the end of the board, she wins a “reward” fish to add to her collection. Winning enough of these rewards unlocks access to the next character.