

Dyscalculia

Introduction

The term dyscalculia refers to an acquired disorder of number processing and calculation skills following brain damage. Henschen was the first to identify this syndrome in 1919.¹ However, for a long time dyscalculia was treated as one of the subcomponents of the Gerstmann syndrome or as an impairment due to more generalised cognitive deficits such as visuospatial and language disorders. It is now well established that impairments in number processing and calculation are independent from deficits in general intelligence, language, reading, writing, semantic memory and short-term memory.²

Acquired deficits in number processing and calculation are rather frequent after brain lesions and may result from both acute and neurodegenerative conditions. The incidence of dyscalculia in patients with either left hemisphere lesions or at the early stage of Alzheimer's disease is high.

Classification

Dyscalculia is not a unitary disorder and can take a variety of different forms. Patients may present with specific impairments in processing numbers, in calculation or in both. Table 1 provides an overview of the types of impairment.

Disorders of number processing

Patients with number processing impairments may show selective deficits in either producing (e.g. reading, writing or repeating) or in comprehending (e.g. knowing that 5 is bigger than 4) Arabic numerals (e.g. 5), verbal numerals (e.g. five) or both.

Patients with number production impairments may show selective deficits affecting syntactical or lexical processing.

- Syntactical errors involve the incorrect selection of the number class to which a number belongs (e.g. they read 600 as "sixty").

- Lexical errors involve the incorrect selection of the position of the individual elements within the correctly identified class (e.g. they read 29 as "forty-nine").

Patients with a deficit in number comprehension may no longer be able to point to the larger of two Arabic numerals (e.g. 345 and 785 or 265 and 2307) or match spoken numerals to the corresponding Arabic numeral. Deficits in number comprehension may selectively affect a subcategory of numbers or different aspects of number meaning. For example, Cipolotti et al³ described a patient who lost the meaning of numbers above 4. Patients with a selective impairment in "cardinal" (which depicts the numerosity of a set of entities and describe the manyness of the set) number meaning or "sequence" (which depicts the position of a number word in the number sequence and do not refer to numerosities) number meaning have been described.⁴

Disorders of calculation

In order to carry out a complex calculation such as 346+475, several independent calculation subprocesses are required. These include:

- The identification of arithmetic symbols (e.g. +, x, -, :)
- The retrieval of arithmetic facts. These are defined as a vocabulary of "number combinations", such as 5+3=8, 3x4=12, 10-4=6. These facts are directly retrieved from memory and the solution to these problems does not require further computational processes or strategies.
- The execution of calculation procedures. These procedures allow access to specific algorithms required to solve multi-digit calculation. Specific examples are the carrying and borrowing procedures.



Natasja van Harskamp studied Neuropsychology and Developmental Psychology at the University of Utrecht. Since 1996 she has been Clinical Neuropsychologist in the Neuropsychology Department of the National Hospital for Neurology and Neurosurgery. She has a special interest in acquired disorders of calculation and social cognition.



Lisa Cipolotti studied Experimental Psychology at the University of Padua and completed a PhD on Dyscalculia in 1993 at UCL. Since 1996 she has been Head of the Neuropsychology Department of the National Hospital for Neurology and Neurosurgery. Her main research is on the acquired disorders of memory, language and calculation.

Table 1 Overview of types of deficits occurring in dyscalculia

Disorders of number processing	Error example	
<i>Disorders of number production</i>	<ul style="list-style-type: none"> • Disorders of syntactical processing (<i>Syntactical errors</i>) • Disorders of lexical processing (<i>Lexical errors</i>) 	<p>5 read as "fifty" 5 read as "seven"</p>
<i>Disorders of number comprehension</i>	<ul style="list-style-type: none"> • Disorders of cardinal number meaning: <i>Problems with identifying the quantity of a set</i> • Disorders of sequence number meaning: <i>Problems with identifying the position of a number in a sequence</i> 	<p>5 is larger than 6 5 comes after 6</p>
Disorders of calculation		
<i>Disorders of arithmetic symbol processing:</i> e.g. adding when there is a multiplication sign		$\begin{array}{r} 47 \\ + 49 \\ \hline 114 \end{array}$
<i>Disorders of arithmetic fact retrieval:</i> e.g. patients failing to retrieve automatically arithmetical facts such as 6x5		
• Operand errors, if the incorrect answer is the correct answer to a problem that shares one of the operands		6x5=25
• Operation errors, if the incorrect answer is the correct answer to another problem involving the same operands, but a different operation		6x5=11
• Table errors, if the incorrect answer is an answer that is a product of two other single digit numbers		6x5=32
• Non table errors, if the incorrect answer is not an operand, table or operation error		6x5=41
<i>Disorders of calculation procedures:</i> e.g. patients failing to apply specific calculation procedures such as:		
• Systematic smaller from larger subtraction errors		$\begin{array}{r} 823 \\ - 644 \\ \hline 221 \end{array}$
• Inappropriate carry over procedures in the calculation of partial products and in the sum of partial products		$\begin{array}{r} 35 \\ \times 97 \\ \hline 245 \end{array}$
		$\begin{array}{r} 171 \\ - 58 \\ \hline 127 \end{array}$
		$\begin{array}{r} 35 \\ \times 723 \\ \hline 245 \end{array}$
		$\begin{array}{r} 275 \\ \times 57 \\ \hline 2995 \end{array}$
		$\begin{array}{r} 385 \\ \times 151 \\ \hline 31001 \end{array}$
<i>Disorders of conceptual knowledge:</i> e.g. the solution of an addition problem is smaller than the addends		8+7=6

Correspondence to:
Natasja J van Harskamp,
Department of Neuropsychology,
National Hospital for Neurology
and Neurosurgery,
2nd Floor, Queen Mary Wing,
Queen Square,
London WC1N 3BG.
Box No. 37
Tel: 0207 829 8793
Email: n.harskamp@ion.ucl.ac.uk
l.cipolotti@ion.ucl.ac.uk

• The retrieval of conceptual knowledge. This allows the understanding of the principles underlying both arithmetic facts and procedures (e.g. the solution of an addition problem is greater than the addends).

Each of these different cognitive processes appears to be functionally independent and differentially susceptible to brain damage.

For example, patients with a selective impairment in the processing of arithmetic symbols misname and misidentify the arithmetical signs and perform the calculation according to their misidentification.⁵

Several patients have been documented with selective arithmetic fact retrieval impairment. Typically, these patients are impaired in solving very simple single digit addition, subtraction, multiplication and division problems. They produce many errors (e.g. $5+7=$ "13 roughly") and their response times are abnormally slow (e.g. >3 sec.). Deficits in arithmetic fact retrieval may be specific for type of operation. Thus, patients have been described with selective impairment or preservation in subtraction, multiplication, addition and division problems (see table 2).

Patients with selective deficits in calculation procedures may have problems in executing those procedures that specify the sequence of steps necessary to solve multi-digit problems, for example, the carrying and borrowing procedures. Patients with a multi-digit subtraction problem may systematically subtract the smaller digit from the larger one regardless of their location in the top or bottom numbers.¹¹ Interestingly, this deficit can be specific for type of procedure. Thus, patients may only have difficulties with the borrowing procedure in complex subtraction, while still being able to carry out complex addition, involving the carry procedure.

Few patients with deficits in conceptual knowledge have been reported. These patients may show a poor understanding of the conceptual aspects of calculation. For example, they produce highly implausible errors (e.g. the solution of subtraction is greater than the minuend) or may not apply very basic arithmetic principles such as order irrelevant-principle in multiplication ($4 \times 12 = 12 \times 4$) or repeated addition of the second operand ($4 \times 12 = 12 + 12 + 12 + 12$).⁸ Selective preservation of conceptual knowledge in the context of severe dyscalculia is also on record.¹² Intact conceptual knowledge may be critical in develop-

ing rehabilitation procedures for arithmetic deficits (see below).

Assessment

A variety of tasks may be used to assess number processing and calculation (see table 3). Recently, a battery of Number Processing and Calculation (NPC) has been standardised by Delazer et al.¹³ Error analyses are also very useful additions to the assessment. They provide invaluable information regarding the type of functional impairment the patient presents.

Localisation

Numerical skills have a discrete and independent brain substrate. The majority of evidence based on lesion studies has indicated the involvement of the left posterior areas. The reports available do not allow for a conclusive localisation of lesions within the left posterior quadrant. However, it appears that the left parietal lobe plays a crucial role. Recent neuroimaging studies have investigated the neuronal correlates underpinning number processing and calculation. They often report large neuronal networks of parietal, pre-

frontal and cingulate areas. In particular, the horizontal segment of intraparietal sulcus bilaterally (HIPS) and the inferior frontal gyrus and the precentral sulcus are mostly implicated.¹⁴

Treatment and recovery

Data on recovery from dyscalculia is rare, however partial recovery occurs in most patients with dyscalculia following vascular lesions.¹⁵ According to the specific type of dyscalculia, different kinds of rehabilitative intervention may be required. Principles of treatment in rehabilitation of dyscalculia mainly consist of:

- reteaching lost arithmetic fact knowledge via extensive training
- errorless learning
- the use of back-up strategies based on the principles underlying arithmetic facts such as the order-irrelevant principle like $8 \times 6 = 6 \times 8 = 48$, decomposition strategies like $4 \times 8 = 2 \times 8 + 2 \times 8 = 32$ or repeated addition of the second operand ($3 \times 5 = 5 + 5 + 5 = 15$).

Case reports show improvement following intense rehabilitation of arithmetic facts and number transcoding deficits.¹⁶

Table 2: Examples of patients with selective impairments/preservations of arithmetic facts

		Multiplication	Subtraction	Addition	Division
Subtraction preservation	BB (Pesenti et al., 1994) ⁶	x	✓	x	-
Subtraction impairment	SS (van Harskamp et al, 2002) ⁷	✓	x	✓	-
Multiplication preservation	JG (Delazer and Benke, 1997) ⁸	✓	x	x	x
Multiplication impairment	VP (van Harskamp and Cipolotti, 2001) ⁹	x	✓	✓	-
Addition impairment	FS (van Harskamp and Cipolotti, 2001) ⁹	✓	✓	x	-
Division impairment	CB (Cipolotti and de Lacy Costello, 1995) ¹⁰	✓	✓	✓	x

✓ = spared; x = impaired; - = not tested

Table 3: Assessment of numerical processing and calculation

Number processing tasks	
Number production	<ul style="list-style-type: none"> • Reading • Writing to dictation • Repetition • Transcoding between Arabic numerals and verbal numerals (5→five)
Number comprehension	<ul style="list-style-type: none"> • Magnitude comparison of Arabic numerals or written/spoken number names (e.g. 12-21) • Analogue number scale task (scale from 0-100, marked at 25; 50; 75, show me 25) • Parity judgment (odd or even number?)
Calculation tasks	
Arithmetic symbol processing	<ul style="list-style-type: none"> • read • point • write the arithmetical signs
Arithmetic fact retrieval	Simple arithmetic problems across the four basic operations such as $4+2$, 3×4 or $5-2$, $6:3$.
Procedural knowledge	Multi-digit calculation, such as $294+12=306$
Conceptual knowledge	Arithmetical principles such as commutativity ($a+b=b+a$); repeated addition; $10a \times 10b$; multiplication/division inversion; $a-1b$; $a+1$; $a-1$; addition/subtraction inversion

References

1. Henschen SE. *Über Sprach-Musik-und Rechenmechanismen und ihre Lokalisationen im Grosshirn.* Zeitschrift für die gesamte Neurologie und Psychiatrie 1919;52:273-98.
2. Cipolotti L and van Harskamp NJ. *Disturbances of number processing and calculation.* In F. Boller & J. Grafman (Eds) *Handbook of Neuropsychology*: 305-31 Amsterdam-New York: Elsevier 2001.
3. Cipolotti L, Butterworth B, Denes G. *A specific deficit for numbers in case of dense acalculia.* Brain: 1991;114:2619-37.
4. Delazer M, Butterworth B. *A dissociation of number meanings.* Cognitive Neuropsychology 1997;14:613-36.
5. Laiacona M, Lunghi A: *A case of concomitant impairment of operational signs and punctuation marks.* Neuropsychologia 1997;35:325-32.
6. Pesenti M, Seron X, Van Der Linden M. *Selective impairment as evidence for mental organization of arithmetical facts: BB, a case of preserved subtraction?* Cortex 1994;30:661-71.
7. Van Harskamp NJ, Rudge P, Cipolotti L. *Are multiplication facts implemented by the left supramarginal and angular gyri?* Neuropsychologia 2002;40:1786-93.
8. Delazer M and Benke T. *Arithmetic facts without meaning.* Cortex 1997;33:697-710.
9. Van Harskamp NJ, Cipolotti L. *Selective impairments in addition, subtraction and multiplication. Implications for the organization of arithmetical facts.* Cortex 2001;38:363-68.
10. Cipolotti L and De Lacy Costello A. *Selective impairment for simple division.* Cortex 1995;31:433-49.
11. Sandrini M, Miozzo A, Cotelli M, Cappa F. *The residual calculation abilities of a patient with severe aphasia: evidence for a selective deficit of subtraction procedures.* Cortex 2003;39:85-96.
12. Hittmair-Delazer M, Sailer U and Benke T. *Impaired arithmetic facts but intact conceptual knowledge - A single case study of dyscalculia.* Cortex 1995;31:139-48.
13. Delazer M, Girelli L, Grana A, Domahs F. *Number processing and calculation-Normative Data from Healthy Adults.* The Clinical Neuropsychologist 2003; 17(3):331-50.
14. Dehaene S, Molko N, Cohen L, Wilson AJ. *Arithmetic and the brain.* Current opinion in neurobiology. 2004; 14:218-24.
15. Basso A, Caporali A, Faglioni P. *Spontaneous recovery from acalculia.* J of the Int Neuropsych Soc 2005;11:99-107.
16. Girelli L. and Seron X. *Rehabilitation of number processing and calculation skills.* Aphasiology, 2001;15(7): 695-12.