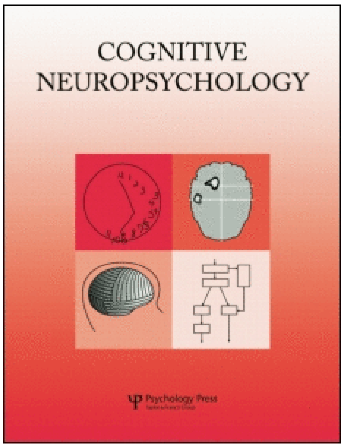

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Dissociations in numerical abilities revealed by progressive cognitive decline in a patient with semantic dementia

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DISSOCIATIONS IN NUMERICAL ABILITIES REVEALED BY PROGRESSIVE COGNITIVE DECLINE IN A PATIENT WITH SEMANTIC DEMENTIA

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This study describes a 3-year follow-up investigation of the deterioration of number abilities in a semantic dementia patient (IH). A few studies have previously reported the decline of number knowledge in patients with degenerative disorders, although almost never in semantic dementia (Diesfeldt, 1993; Girelli, Luzzatti, Annoni, & Vecchi, 1999; Grafman, Kempen, Rosenberg, Salazar, & Boller, 1989). These studies described the change of the patients' performance mainly in terms of increased errors in number tasks. On the other hand, dissociations between different types of number abilities, or different arithmetical operations, have been reported in patients with focal lesions. In the present investigation, the cognitive basis of number processing was revealed throughout the patient's cognitive decline. Two major results emerged from a longitudinal study: First, the patient's conceptual knowledge of arithmetic was well preserved despite severe impairment of nonarithmetic conceptual knowledge. Second, the patient's progressive decline revealed patterns of dissociations between different number abilities. These were between (1) multiplication and other arithmetical operations, which particularly emerged in the use of algorithms; (2) impaired knowledge of number facts and procedures on one hand, and conceptual knowledge of arithmetic on the other; and (3) different types of transcoding skills. The implications of these dissociations for the cognitive architecture of number processing are discussed.

INTRODUCTION

It is now established that numerical knowledge can survive when other conceptual or semantic knowledge has been lost (Cappelletti, Butterworth, & Kopelman, 2001; see also Crutch & Warrington, 2002). This paper focuses on the deterioration of numerical knowledge in the absence of other knowledge. This provided a good opportunity to dissect numerical knowledge into its subcomponents. Previous studies on the deterioration of number knowledge mainly described an increase

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of errors in number tasks, especially calculation. Other more detailed investigations of the dissociation between number abilities have been reported, although they did not explore the patients' performance over time. In the following, both cross-sectional and longitudinal studies of number knowledge will be presented, which revealed different subcomponents of numerical processing.

Decline of number knowledge

Only three studies have reported the deterioration of numerical knowledge in patients with neurodegenerative disorders. The first of these studies described a patient with probable Alzheimer dementia (GC) whose number skills were tested with four different test batteries (Grafman et al., 1989). The patient initially showed impaired arithmetical procedures and defective retrieval of simple facts when performing multiplication and division problems. Simple number tasks, such as magnitude comparison, were relatively better preserved. In a follow-up investigation, the patient's performance declined in simple number tasks and in calculation.

The second study described a follow-up investigation of number skills in a semantic dementia patient (Diesfeldt, 1993). The patient could read and write numerals and number words to dictation, and performed simple and multidigit arithmetical operations well, with the exception of multiplication tables, which declined significantly over the course of the illness. Third, a very short report described the decline of numerical skills in a patient with probable Alzheimer dementia (Girelli, Luzzatti, Annoni, & Vecchi, 1999). In this patient, both arithmetical procedures and number facts were impaired (although number facts less so). In these studies, it was not clear what the contribution of general cognitive deterioration was to the reported declines in mathematical abilities.

Selective deficits and sparing of number knowledge

Several neuropsychological studies have shown dissociations between various numerical abilities in patients with nonprogressive disorders, although the patients' performance was not considered over time. For the purposes of the present study, patterns of selectively impaired and spared performance will be considered in conceptual knowledge of arithmetic and between different arithmetical operations.

Types of knowledge of arithmetic

We need to distinguish between a number of terms that we will use.

Number facts are items that have been learned by rote. This is especially true of the multiplication tables, also called "times-tables" ($N \times M$, e.g., 2×4), but will also be the case for single digit (e.g., $2 + 4$) and some other addition facts (e.g., $11 + 11$).

Arithmetic rules are the universal $n + 0 = n$; $n \times 1 = n$; $n \times 0 = 0$.

Arithmetic procedures are the individual steps that have to be taken during calculation, such as starting at the rightmost digits in addition, carrying, and so on. There are *canonical* procedures—those taught in school (see also Figure 3).

Algorithms is the term we use for noncanonical procedures.

Conceptual knowledge consists of "an understanding of arithmetical operations and *laws* pertaining to these operations" (Hittmair-Delazer, Semenza, & Denes, 1994, p. 717; also Hittmair-Delazer, Sailer, & Benke, 1995; Sokol & McCloskey, 1991). This understanding would include the commutative law, (e.g., $3 \times 5 = 5 \times 3$, or $3 + 5 = 5 + 3$), the distributive law (e.g., $[(5 + 3) \times 2] = [(5 \times 2) + (3 \times 2)]$), and the associative law. Arithmetical operations can be solved on the basis of these principles, when arithmetic facts cannot be recalled from memory (e.g., patients BE and DA, Hittmair-Delazer et al., 1994, 1995). Conceptual knowledge has not been exhaustively considered in cognitive models of number processing (Dehaene & Cohen, 1995; McCloskey, Caramazza, & Basili, 1985).

The understanding of arithmetical operations and the use of arithmetic principles (rules or laws) underlying them can be selectively impaired or spared. On the one hand, conceptual knowledge dissociates from simple fact retrieval and from the ability to use arithmetical procedures.

For example, patients DRC (Warrington, 1982), BE (Hittmair-Delazer et al., 1994), MW (McCloskey et al., 1985), IE (Sokol et al., 1989), and GE (Sokol, McCloskey, & Cohen, 1989; Sokol & McCloskey, 1990, 1991) used back-up strategies, which we will call algorithms, to compensate for their impairment with simple facts and/or with arithmetical procedures. On the other hand, the selective impairment of conceptual knowledge has also been reported in the context of preserved knowledge of arithmetical facts (Delazer & Benke, 1997). It is worth noting that although some patients have been reported to use algorithms instead of canonical procedures to solve arithmetical operations, this is not routinely observed in patients with numerical impairments.

Other studies that reported selective impairment of arithmetical facts or procedures gave no indication of the patients' understanding of the operations they could not process. For instance, patient BBO (Dehaene & Cohen, 1997) had severe problems in solving single-digit multiplication problems, including very simple ones (e.g., 2×3). However, there was no independent indication of the preservation of conceptual knowledge.

Although conceptual knowledge of arithmetic dissociates from factual and procedural knowledge, the relation between arithmetical and nonarithmetical conceptual knowledge has not yet been studied. Patients with semantic dementia are ideal candidates for exploring this issue. Their understanding of arithmetical concepts can be compared and contrasted with their understanding of nonarithmetical concepts. This issue has not previously been explored when patients with semantic disorders were investigated (e.g., Crutch & Warrington, 2002; Grafman et al., 1989).

Dissociation between arithmetical operations

Another type of dissociation that has been frequently reported in patients with nondegenerative disorders is between arithmetical operations. Their functional and anatomical architecture has been differently explained in various theoretical accounts. The most influential one is perhaps Dehaene and Cohen's Triple Code model (Dehaene & Cohen, 1995), which suggested that "dissociations between operations (. . .) reflect the underlying structure of the two main cerebral pathways for calculation" (Dehaene & Cohen, 1997, p. 243), these pathways being rote verbal and quantitative knowledge of arithmetic. In particular, the authors observed that there is often a selective sparing of subtraction problems as opposed to multiplication and addition (Dagenbach & McCloskey, 1992; Dehaene & Cohen, 1997; Lampl, Eshel, Gilad, & Sarova-Pinhas, 1994; McNeil & Warrington, 1994; Pesenti, Seron, & Van der Linden, 1994). The model also predicted that there would be a relationship between subtraction and general quantity manipulation.

More recent evidence has challenged Dehaene and colleagues' position. In particular, Van Harskamp and colleagues (Van Harskamp & Cipolotti, 2001; Van Harskamp, Rudge, & Cipolotti, 2002) presented a patient (FS) with selective impairment of addition problems and preserved subtraction and multiplication, and another one (DT) with a selective deficit in subtraction problems with intact quantity manipulation. This debate on the nature of the dissociations between arithmetical operations motivates further explorations in patients with a degenerative disorder. The nature of their disease makes the investigation especially interesting. Changes over time in performing arithmetical operations can provide information about the underlying cognitive structures specific for single operations.

Aims of the present study

It appears that previous studies explored either the deterioration of number knowledge in terms of the increase of errors in number tasks, especially calculation, or the dissociation between numerical abilities at a given time. The current study aimed to: (1) investigate the cognitive basis of number processing throughout the patient's cognitive decline; and (2) explore the nature of arithmetical conceptual knowledge.

Arithmetical conceptual knowledge can be considered a high level number semantic skill. Other numerical competences, such as counting, reading numbers aloud, or answering numerical questions do not seem to require the same advanced level of comprehension or manipulation of numerical concepts and conceptual knowledge does. This study aims at finding out to what extent a high-level semantic process such as arithmetical conceptual knowledge is preserved in the context of severe semantic impairment. In order to achieve these goals, we investigated the decline of number and calculation skills in a patient (IH) with semantic dementia.

CASE REPORT

When initially seen (1998), IH was a 64-year-old, right-handed, former banker with 12 years of formal education. He showed preserved general intelligence measured with nonverbal tasks; he had severe comprehension and naming difficulties but relatively well-preserved knowledge of very familiar topics, such as sports and political events, as well as explicit memory for everyday events. The details of IH's neuropsychological background tests and MRI scan have been reported in a previous paper (Cappelletti et al., 2001). Table 1 shows a summary of the patient's performance in general neuropsychological and semantic tests. Despite his severe semantic impairments, IH seemed to be able to use numbers appropriately in everyday life: He could use money, tell the time and the date, and use numbers to gamble and play the lottery.

An MRI brain scan with coronal slices showed very severe disproportionate left temporal lobe atrophy. There was relative sparing of the left hippocampus, but it did show some atrophy, and there also appeared to be some minimal widening of the subarachnoid space surrounding the right temporal lobe, implying a much lesser degree of atrophy. There was no discernable change in the appearance of the scans between 1995 and 1998.

EXPERIMENTAL INVESTIGATION

The data reported here were collected over approximately 3 years, from October 1997 to August 2000. Only IH's numerical knowledge will be reported as his non-number semantics have already been shown to be severely impaired elsewhere. Part of the patient's performance on number tasks has previously been reported (Cappelletti et al., 2001). Here we will describe IH's number skills over time, focusing on the strategies he developed to perform numerical tasks, in particular arithmetical operations. This will allow us to explore in more detail the nature of arithmetical conceptual knowledge.

Methods and materials

A comprehensive description of the materials and the procedures used to test IH's number knowledge has already been reported (see Cappelletti et al., 2001; and Appendix A for a more detailed description of the tests used). The number battery was composed of a nonverbal and a verbal section, the latter including number and calculation tasks respectively. Tasks assessing nonverbal number knowledge consisted of number processes that are not primarily a product of verbal abilities. The amount of linguistic resources required to perform these tasks is minimal. On the other hand, a large proportion of number tasks depend on or are by-products of general verbal abilities. These tasks are distinguished between those requiring a simple manipulation of numbers or quantities (e.g., dot enumeration, counting, and number transcoding), and those requiring the application of arithmetical procedures, the recall of arithmetical facts, or the ability to approximate to a result.

Numerical tasks where IH failed were not repeated in subsequent examinations (e.g., approximation tasks, see Appendix A). In addition, as informal observation suggested that the patient's attention and concentration abilities seemed to be slightly impaired, some tasks were adjusted accordingly. For instance, the Graded Difficulty Arithmetic Test (Jackson & Warrington, 1986) was presented in written format, while the experimenter read the operations aloud, although it is usually just presented orally. As the task itself is demanding, a written presentation of the stimuli would partially reduce the cognitive load, and allowed us to distinguish short-term memory or concentration disorders from difficulties in solving the number task itself. IH's performance will be

Table 1. *Control subjects and IH's performance on general neuropsychological and semantic tasks (per cent or number correct)*[a]

	Testing time-periods			
Tasks performed	*1996*	*1998*	*1999*	*Controls*
General intelligence				
Raven's Coloured Progressive Matrices[b]	72.2%	88.9%	69.4%	50th–75th %ile
Language				
Reading aloud: Regular words	92%	76%	32%	Pattern of surface dyslexia
Irregular words	74%	40%	15%	
Memory				
Logical Memory Test I + II			0	
Recognition Memory Test[d]: Words	52%		0%[k]	
Faces	82%		52%	
Executive functions				
Cognitive Estimate			Not understood	
Modified Card Sorting Test[c]	6/6 categ		6/6 categ	Normal
Semantic tasks				
Verbal tasks				
Graded Naming Test[e] ($N = 30$)	40%		0%	IQ < 76, 63
Picture naming[f]	40%	8%	0%	
Category naming ($n = 40$)			25%	99
Naming real objects ($n = 15$)			0%	100
Word classification ($n = 50$)			0%	100
Name-to-picture matching ($n = 40$)	56%		55%	97
Pyramid and Palm Tree[i] (verbal, $N = 52$)			0%[k]	99
Phonological fluency (FAS)	8 items		0 items	42[l]
Semantic fluency (total 8 categories)			0 items	117[l]
Verbal definition ($n = 73$)			0%	99.5
Pictorial tasks				
Picture classification ($n = 40$)			80%	99
Subcategory picture classification ($n = 9$)			66%	100
Size judgement task ($n = 20$)			65%	99
Object decision task ($n = 20$)			70%	88
Pyramid and Palm Tree[i] (pictorial, $N = 52$)	86%		52% (chance)	99

[a]Adapted from: Cappelletti, M., Butterworth, B., & Kopelman, M. (2001). Spared numerical abilities in a case of semantic dementia. *Neuropsychologia, 39* (11), 1224–1239.

[b]Raven (1965); [c]Nelson (1976); [d]Warrington (1984); [e]McKenna and Warrington (1983); [f]Snodgrass and Vanderwart (1980); [g]Benton and Hamsher (1976); [h]Kay, Lesser, and Coltheart (1992); [i]Howard and Patterson (1992); [j]Wechsler (1987).

[k]0% score indicates that the patient could not engage in the task at all.

[l]Mean items produced.

analysed on the basis of the main numerical skills; when appropriate, the results will be divided into different time-periods according to the occurrence of the main changes in performing those tasks.

Nonverbal number tasks

IH performed very well on nonverbal number tasks during all the investigation times, except for the last (i.e., August 2000). Table 2 shows the patient's performance in numerical and calculation tasks. For about 3 years following the first examination (October 1997–March 2000) IH showed good ability in ordering dots and numbers from the smallest to the largest set, in comparing quantities and composing the value of numbers with tokens (see Appendix A). Together with number comparison, the ability to compose the value of numbers

Table 2. *Control subjects' and IH's performance on number and calculation tasks (per cent correct and SD)*

Tasks performed	*Controls*	*IH*			
Nonverbal number tasks		*Oct1997–Mar2000*	*Aug2000*		
Dot seriation ($N = 18$)	100	100	NT		
Dot magnitude comparison ($N = 18$)	100	100	Impossible[a]		
Number seriation ($N = 18$)	100	100	NT		
Magnitude comparison ($N = 20$)	99.4 (1.7)	100	Impossible		
Number composition with tokens ($N = 48$)	100	99.3	82; impossible		
Placing numbers on an analogue line ($N = 36$)	100	100	Impossible		
Verbal number tasks		*Oct1997–Mar2000*	*Aug2000*		
Number recognition					
Spoken number words to numerals ($N = 18$)	100	98	NT		
Numerals to spoken number words ($N = 18$)	100	100	NT		
Dot enumeration ($N = 10$)	100	100	100		
Counting ($N = 40$)	100	100	100; 28.5[b]		
What comes next/before ($N = 40$)	100	100	Impossible		
Bisection task: Numbers ($N = 10$)	100	90	10		
Letters/Months/Days ($N = 30$)	100	100/96/6.2.5	0		
Knowledge of number facts: Personal ($N = 10$)	100	5	NR		
Nonpersonal ($N = 10$)	100	0	NR		
Transcoding		*Oct97–Mar99*	*Mar99–Mar00*	*Aug2000*	
Reading					
One to four-digit numerals ($N = 135$)	100	98	98	See Table 3	
Five-digit numerals ($N = 10$)	98.4 (2.1)	100	90		
Six-digit numerals (N = 10)	96 (1.8)	100	50		
Number words ($N = 50$)	100	100	100		
Writing to dictation					
One to four-digit numerals ($N = 100$)	100	100	100; 89	See Table 3	
Five-digit numerals ($N = 10$)	98 (1.4)	90	30		
Six-digit numerals ($N = 10$)	98 (1.6)	90	—		
Number words ($N = 35$)	100	100	100		
6 → SIX ($N = 20$)	100	100	12.5	See Table 3	
SIX → 6 ($N = 20$)	100	100	88.5	See Table 3	
Calculation tasks		*Oct97–Aug98*	*Oct98–Feb99*	*Mar99–Mar00*	*Aug00*
Mental calculation (single-digit operations)					
Addition problems ($N = 200$)	100	98	100	98	73
Subtraction problems ($N = 154$)	100	95	100	95	NT
Multiplication problems ($N = 239$)	90 (6.8)	73[c]	84	73	NT
Division problems ($N = 50$)	88 (8.2)	NT	66	75	NT
Written calculation (multidigit operations)					
Addition problems ($N = 128$)	99.4 (0.6)	99	98.7	96	NT
Subtraction problems ($N = 64$)	98 (2.4)	96	99.3	93	NT
Multiplication problems ($N = 42$)	95 (6.6)	69	55	48	NT
Division problems ($N = 40$)	95 (7)	62	66	71	NT
Graded Difficulty Arithmetic Test[d] ($N = 28$)		96[e,f]	75[f]	NT	32[f,g]
Approximation to the correct result ($N = 12$)	100	0[h]	NR	NR	NR

[a] When performance indicated as impossible, IH did not attempt the task, making no response at all.
[b] Results of a second testing session in August 2000.
[c] 100 single-digit multiplication operations administered at this time.
[d] Jackson and Warrington (1986).
[e] Scaled-score corresponding to superior level.
[f] Performance scored without the prescribed timing criteria.
[g] Scaled-score corresponding to dull average.
[h] Instructions not understood.
NT = Not tested; NR = Not retested.

is an important measure of the capacity to understand and manipulate quantities. IH had no difficulties in placing numbers along a line. During the last examination, the patient was severely impaired in performing number tasks possibly because of serious comprehension problems that interfered with testing. IH's knowledge of number sequence was impaired, unless dots were used. Number comparison could not be performed again because of problems in understanding task instructions. Number composition with tokens was relatively preserved at least for two-digit numbers.

Verbal number tasks

For about 3 years following the first examination (October 1997–March 2000), IH performed well on verbal number tasks. He could recognise spoken and written numbers, count and enumerate dots, indicate what number comes before or after a given one or between two numbers (with only a few errors in the latter task). His performance changed dramatically at the time of the last examination (i.e., August 2000), where IH showed severe impairments in performing many numerical tasks. As with the nonverbal number tasks, it is possible to suggest that the patient's comprehension disorders interfered with his ability to perform cognitive tasks including numerical ones.

The patient's performance in transcoding tasks will be examined in detail. Results are divided in different time-periods, which are clearly defined, according to the occurrence of the most significant changes in performance.

Transcoding

Reading numerals and number words. For the first 2 years of investigation (October 1997–March 1999), IH's performance on reading and writing numerals and number words was very well preserved. He was able to read aloud numerals and number words up to six digits and to write them to dictation or from another numerical format (e.g., two from the spoken TWO or the written 2).

When subsequently examined (i.e., after March 1999), the patient continued to perform well with one- to four-digit numbers (only 8 mistakes out of a total of 346 items presented, 2%) and at ceiling with number words (100% correct answers out of 116 trials). However, he made errors in reading multidigit Arabic numbers, especially six-digit numerals. The 9 errors made (out of 10 trials) were 1 lexical, 1 omission, and 1 mixed error, in addition to 6 other syntactic mistakes.[1]

During the last examination (i.e., August 2000), IH performed at ceiling in reading aloud single and two-digit numerals, but made phonological errors in reading written number words (e.g., six was "son", then "sixty", and finally "six"; THIRTY-FIVE was "Thursday-Friday"). In addition, when reading three- and four-digit numbers, IH consistently said "hospital" for hundred and "Thursday" for thousand whether numerical stimuli were presented in Arabic or verbal format.

Writing numerals and number words. For over 2 years following the beginning of the examination (October 1997–November 1999), IH performed well in writing one- to four-digit numerals to dictation and in writing numerals from written number words (i.e., ONE → 1, 111 out of 116 correct answers, 96%).

During subsequent examinations (i.e., after November 1999) IH's performance changed significantly, McNemar Test, $\chi^2(1) = 11.1$, $p < .001$; see Table 1. In writing numerals to dictation, errors were mainly made with four-digit numerals, especially with those containing or ending in zero (8 out of 13, 61%). For instance, IH wrote "eight thousand eight hundred and twenty" as 820. No mistakes were made with single and teen numerals and those occurring with two- and three-digit numerals were all lexical. Writing five-digit numerals to dictation seemed even more problematic as IH wrote correctly only 3 out of 10 numerals. Errors

[1]Following Seron and Deloche (1984), transcoding errors were classified as lexical (e.g., "9" read as eight instead of nine), syntactic (e.g., "9" read as nine thousand instead of nine), and lexical-syntactic (e.g., "7110" read as five hundred and eleven instead of seven thousand one hundred and ten).

were syntactic, lexical, mixed, and omissions. For instance, “forty thousand and twenty-five” was written as 40250 and “thirty-nine thousand” was written as 13000.

Table 3 summarises the patient’s performance on transcoding tasks during the last examination (i.e., August 2000).

At this time, IH could not write number words to dictation, irrespective of the type of number. All the target number words were written as numerals (e.g., 2 instead of TWO, or 15 instead of FIFTEEN), although some attempts to write them in verbal format were made (“eight” was written as EIGHT). The only number word IH managed to write was hundred, e.g., “four hundred” → 4 HUNDRED. Writing single numerals to dictation was preserved, and teens were written in a way that the first spoken part corresponded to the first digit, followed by another number. This number was usually 1 (e.g., “*four*teen” → *4*-1) and sometimes another digit (e.g., “*six*teen” → *6*-9). However, when retested a few weeks later the patient consistently produced the number 0 (e.g., “thirteen” → 30, “fifteen” → 50). In writing three- and four-digit numbers, IH consistently made two types of mistakes: complete lexicalisation errors from oral input, and partial lexicalisation errors from written input.[2] It seems possible to suggest that IH’s impairment in transcoding may be explained in terms of selective phonological difficulties. We discuss this possibility in greater detail in a later paper (Cappelletti, Morton, Butterworth, & Kopelman, 2004b).

During the last examination, IH’s performance also changed in writing numerals from number words and vice versa. In the first task (i.e., ONE → 1), IH performed relatively well, although significantly worse than before, McNemar Test, $\chi^2(1) = 15.06$, $p < .001$. Errors were syntactic and consisted of inserting an extra zero. For instance, “two hundred and twenty” was written as 2020 and “three hundred and twenty one” as 3021. In the second task (i.e., 1 → ONE), IH was even more impaired. He only gave six answers and refused to continue the task after that. Only two answers were correct, and three of the errors consisted of reproducing the answer in the same format as the input, e.g., 11 → 11. Another error consisted of a mixture of numerals and letters, namely 4 was transcoded as 4NE (note that none of the letters produced corresponded to the target number name, i.e., four). Although not explicitly asked to do so, IH read all the 16 numbers correctly.

Conclusion on IH’s performance on transcoding tasks. During the first 2 years of examination, IH performed well on reading and writing one to four-digit numerals and number words to dictation or from another number format. When subsequently tested, the patient made errors with large numbers

Table 3. *IH’s performance on transcoding tasks at time-period 4 (August 2000)*

Stimuli	*‘1’ to dictation*	*‘one’ to dictation*	*Reading ‘1’*	*Reading ‘one’*	*‘1’ → ‘one’*	*‘one’ → ‘1’*
Single-digit	9/9	Impossible[a]	9/9	7/7	4/4	Impossible
Teens	4/9, 2/9[b]	Impossible	4/5	4/5	3/4	Impossible
Two-digit	12/12	1/6	2/4	1/5	3/3	Impossible
Three/four-digit	1/17	0/5	9/9	8/8	3/8	Impossible
Five/six-digit	NT	NT	0/20	NT	NT	NT

[a]When performance indicated as impossible, IH did not attempt the task making no response at all.
[b]Results of two different testing sessions.
NT = Not tested.

[2] Complete lexicalisation errors consist of producing all the elements that compose a multidigit number in Arabic format, namely the unit of multipliers (i.e., *1*00 for “hundred”), and the zeros (i.e., 1*00* for “hundred”) e.g., “six hundred” → 6100 (Noel & Seron, 1995). Partial lexicalisation errors consist of producing only the zeros of a multiplier but not the unit, e.g., SIX HUNDRED AND TWO → 6002 (Macoir, Audet, & Breton, 1999; Noel & Seron, 1995).

and showed a dramatic decline in the performance during the last examination: some transcoding tests could no longer be executed (e.g., writing number words to dictation), many errors were made especially with complex numbers (in terms of amount of digits or structure). Errors were syntactic, lexical, perseverations or omissions.

Calculation tasks

IH's performance on calculation tasks will be analysed in terms of single- and multi-digit operations. The first consists of orally presented problems comprising digits from 0 to 9 and requiring the patient to produce an oral answer. Multidigit operations consist of problems with two- to four-digit operands presented in written format and requiring the patient to produce a written answer. Another task with multidigit problems was also administered to IH (Graded Difficulty Arithmetic Test; Jackson & Warrington, 1986).

The patient's performance on calculation tasks with single- and multi-digit operations is divided into approximately four time-periods according to the times when the most significant changes in performance occurred. In the following, IH's performance on calculation tasks will be described in detail, and the different time-periods are clearly defined.

Single-digit operations

Time-period 1: October 1997–August 1998. When first tested, IH could solve single-digit operations quite well, although a few errors were made in multiplication problems. Towards the end of time-period 1, IH's performance changed, and in the following section it will be analysed in detail.

The answers to single-digit arithmetical problems orally presented were classified as fact retrieval when the patient produced them spontaneously and relatively quickly, omission when no answer was given, and strategy when the answer resulted from the application of an algorithm (Hittmair-Delazer et al., 1994). Initially, IH performed single-digit multiplication problems by retrieving the answers from memory. Overall, he made 27% of errors, 18% in fact-based problems (i.e., $N \times M$) where at least one operand was 8 or 9, and 9% in rule-based problems (i.e., $N \times 0$, $N \times 1$).

Subsequently, results of operations were obtained by using a specific algorithm (referred to as Algorithm 1). Taking the operation 8×6 as an example, Algorithm 1 consists of selecting one of the operands, for example 6, and mentally multiplying it by 2 (i.e., $6 \times 2 = 12$). The result is added up as many times as required to reach the other operand (i.e., 4 in this case, $12 + 12 + 12 + 12 = 48$). Similarly, in the operation 8×9, IH selected the operand 9, multiplied it by 2 (i.e., 18), and added 18 as many times as indicated by half of the other operand (i.e., 4). An example of the use of this algorithm is given in Figure 1.

Algorithm 1 was used in 7% of the operations and always led to correct results. It was only employed when both the operands within an operation were bigger than 5 (the only exception being 6×6). It seems therefore that the size of the operands induced IH to apply algorithm 1, presumably because the 6 to 9 times tables were not available to him. Algorithm 1 was based on knowledge of simple additions (i.e., $N + M$), of rules (i.e., $N + 0$), of the two times table (i.e., $N \times 2$), and of the commutative law, namely the principle that the result of 6×8 is the same as that of 8×6. More critically, IH also indicated that he could understand the complementarity of multiplication and division, namely that multiplying M by N and dividing it by N does not change the value of M (for instance, that $8 \times 9 = [(8/2) \times (2 \times 9)] = 4 \times 18$). IH developed and applied this strategy spontaneously when operations where presented orally, wrote the numbers in a column, and recited them aloud while solving the problems.

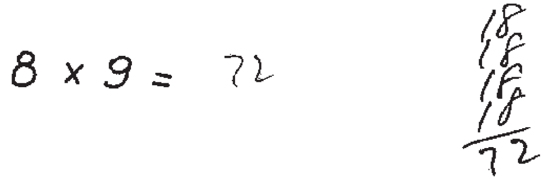

Figure 1. *An example of the use of Algorithm 1 in IH's performance.*

Time-period 2: October 1998–February 1999. During time-period 2, IH performed at ceiling on addition and subtraction problems, and solved correctly 201 out of 239 multiplication problems (84%). This seems to suggest that his knowledge of single-digit multiplication tables was still relatively well preserved. However, a deeper analysis of IH's performance showed that he did not recall simple tables from memory as much as before, and that he used algorithm 1 significantly more (7% vs 37%), McNemar Test, $\chi^2(1) = 46.02$, $p < .001$, and more inefficiently (31% errors compared to no errors at time-period 1). Algorithm 1 was also employed to solve multiplication problems with small operands (i.e., smaller than 5, e.g., 2×7 was solved as $7 + 7$), whereas at time-period 1 it was only used with big operands.

The patient also spontaneously developed and used another algorithm (Algorithm 2), which was mainly employed with big operands. Once more, it seems that the size of the operands induced the use of this algorithm. Algorithm 2 consists of selecting the first operand of the problem (i.e., the leftmost, for instance number 5 in 5×4), and of reciting the table corresponding to it as many times as indicated by the other operand (e.g., 4 in the above example, that is 5, 10, 15, 20). The sequence ends when the table corresponding to the second operand is reached (e.g., 20).

Figure 2 shows the proportion of single-digit multiplication recalled from memory or solved with Algorithms 1 and 2 during time-period 2.

Errors made in solving multiplication problems were classified as follows: (1) operand errors, e.g., $2 \times 4 = 12$, were those corresponding to the result of a different multiplication problem; (2) non-table errors, e.g., $2 \times 4 = 13$, were those that do not exist in the multiplication tables; (3) rule errors were those occurring in performing operations involving 0 and 1, such as 4×0 or 1×4; (4) omissions, when no result was produced (McCloskey, Aliminosa, & Sokol, 1991). Table 4 indicates the type and proportion of errors made in multiplication problems.

Overall, IH's performance on single-digit multiplication problems indicates a decline from time-period 1 in terms of the strategies used: Simple

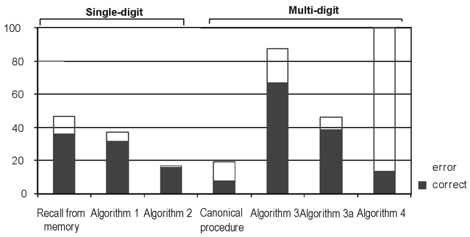

Figure 2. *Single-digit multiplication operations recalled from memory or by using Algorithms 1 and 2 (per cent of use), and multidigit multiplication operations[a] solved with Algorithms 3, 3a, and 4 at time-period 2. The dashed areas at the top of each column indicate the proportion of errors made within each strategy used.*

[a]*Considering the total number of operations an algorithm could be applied to.*

facts were not just recalled from memory but also performed through other procedures.

Time-period 3: March 1999–March 2000. At time-period 3 of the experimental investigation, IH performed well on addition and subtraction problems, which were solved with very few mistakes (see Table 1). Errors were always very close to the correct result. For instance, the result of $9 + 6$ was 17 instead of 15. Division problems were performed with some errors (25%) occurring in problems with the rule $n \div 1 = n$. The same type of error was also produced at time-period 2. It seems, therefore, that for these types of operations there was no difference between time-period 3 and the previous investigation.

Conversely, performance on multiplication problems showed a significant decline from the previous assessment, McNemar Test, $\chi^2(1) = 33.03$, $p < .001$. Table 4 shows the type and the proportion of the errors made. Results indicated that: (1) errors mainly occurred in solving problems with rules (i.e., $N \times 0$, $N \times 1$); (2) towards the end of the examination, errors emerged in operations with small operands (smaller than 5); initially, errors had only been made in operations with big operands; (3) the same procedures described at time-period 1 and 2 were still employed at time-period 3 and no new procedures were used.

Table 4. *Classification of errors in performing single and multidigit multiplication operations at time-periods 2 and 3 (number of errors, and in brackets per cent of errors over the total number of errors for each type of operation)*

	Time-period 2 (Oct98–Feb99)	*Time-period 3 (Mar99–Mar00)*
Single-digit operations	Total $N = 239$	Total $N = 321$
Operand	7 (17.9)	13 (15.1)
Non-table	8 (20.6)	21 (24.4)
Rules	21[a] (53.8)	51 (59.3)
Omissions	3 (7.7)	0
Other	0	1 (1.7)
Total errors	39	86
Multidigit operations	Total $N = 52$	Total $N = 62$
Part of procedure missing or wrong[b]	17 (40.4)	14 (46.6)
Wrong adding up	11 (26.3)	7 (23.4)
Wrong partial product	14 (33.3)	3 (10)
Part of result missing	–	4 (13.3)
Other	–	2 (6.7)
Total errors	42	30

[a]Out of 35 rule-based problems presented.

[b]For example, the wrong operand was multiplied (e.g., in the operation 57×48, 5 is multiplied by 7). Note that this is a procedural error in the context of a procedure applied to the correct arithmetical operation.

Time-period 4: August 2000. At time-period 4 of the experimental investigation, IH's cognitive decline and severe comprehension difficulties did not allow extensive examination of his calculation skills. The patient could only be tested on single-digit addition problems that he performed relatively well (73% correct answers).

Conclusions on IH's performance on single-digit operations. IH's performance during approximately 3 years of investigation showed progressive decline in the ability to perform single-digit arithmetical operations. This decline did not affect arithmetical operations equally: While addition and subtraction problems were relatively preserved, performance in multiplication operations showed an increased number of errors and the use of strategies that the patient employed to overcome the loss of memorised tables. Division problems were also impaired, although the patient was able to use some alternative strategies to solve them. IH's ability to spontaneously develop and use compensatory procedures to perform calculation strongly suggests that he could understand and manipulate the principles underlying arithmetical operations. Note that IH always applied algorithms appropriately, that is to the arithmetical operations they could legitimately be applied to. For example, Algorithm 1 was only applied to multiplication and not to subtraction or addition problems.

Multidigit arithmetical problems

Time-period 1: October 1997–August 1998. When initially tested, IH could solve multidigit addition and subtraction problems almost at ceiling, although a few errors were made with multiplication problems, $\chi^2(1) = 7.86$, $p < .01$, and $\chi^2(1) = 3.24$, $p < .10$, respectively. Errors consisted of wrong alignment of digits or of errors in adding up the partial products. Multiplication problems were always performed with a canonical procedure. Figure 3 illustrates the steps that compose a canonical procedure, and Figure 4 gives an example of IH's performance based on it.

IH performed very well on multidigit operations of the Graded Difficulty Arithmetic Test (Jackson & Warrington, 1986). When his performance was

23×		
35=		
115	→	1st partial product. (5×3)=15, 5 written, 1 carried. (5×2)=10+1 carried=11
69–	→	2nd partial product. Space below the rightmost digit. (3×3)=9, (3×2)=6
805		Sum of partial products. Carrying required.

Figure 3. *The steps of a canonical procedure used to solve multidigit multiplication problems. The operation 23 × 35 is taken as an example.*

scored without the prescribed timed criteria, it corresponded to superior level. The patient could not perform an approximation task, possibly because of difficulty in understanding the complex instructions. This test was not repeated in subsequent examinations.

Time-period 2: October 1998–February 1999. At this time, IH could still solve multidigit addition and subtraction problems very well. On the other hand, his performance with multiplication operations showed that: (1) the proportion of errors increased significantly, McNemar Test, $\chi^2(1) = 8.1$, $p < .01$; see Table 1; (2) canonical procedures were no longer used even when the patient was encouraged to use them; (3) other algorithms were employed instead.

An algorithm based on the associative law was introduced to solve 2 × 2-digit multiplication problems (Algorithm 3). Algorithm 3 consists of selecting one of the operands, e.g., 35 in the operation 23 × 35, and in multiplying it by 10 (i.e., 350). This number is then rewritten as many times as indicated by the decade of the other operand. For example, for the decade 2 of 23, 350 is rewritten twice, e.g., 350, 350 (A). Subsequently, 35 is multiplied as many times as the unit of the other operand (e.g., 3 in 23, therefore $35 \times 3 = 105$), and the result is indicated as (B). Finally, (A) and (B) are added together, e.g., $350 + 350 + 105 = 805$. Algorithm 3 consists of five steps, is based on the ability to perform multidigit addition problems, on the knowledge of the 2 times table, and on the ability to multiply numbers by 10. More critically, it is based on the rule stating that within an operation operands can be grouped without changing the result, for instance $18 \times 15 = [(18 \times 10) + (18 \times 5)]$. Figure 5 shows an example of an operation performed with Algorithm 3.

A slightly modified version of Algorithm 3 (Algorithm 3a) was used when the unit of one of the operands consisted of numbers 9 or 1, (e.g., the operations 19 × 17 or 21 × 28). These operands were treated as decades, and the value of the other operand was added or subtracted accordingly. For instance, the operation 19 × 17 was treated as $[(20 \times 17) - 17]$. In particular, 17 is first multiplied

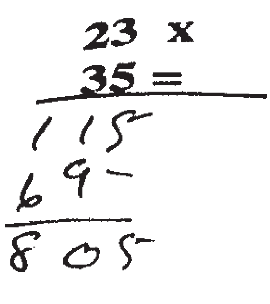

Figure 4. *An example of IH's performance in solving a multidigit multiplication operation using a canonical procedure.*

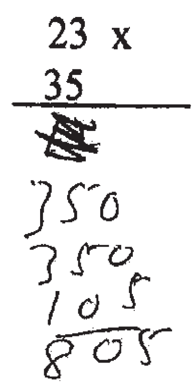

Figure 5. *An example of IH's performance in solving a multidigit multiplication problem using Algorithm 3.*

by 10 (A), and then rewritten as many times as indicated by the decade that follows that of the other operand (i.e., *1*9), in this case, 2, therefore 170, 170. All the rewritten items are then added up, e.g., in this case $170 + 170 = 340$, and indicated as (ATot). Finally, 17 is subtracted from (ATot), e.g., $340 - 17 = 323$. Algorithm 3a consists of five steps, and is based on the ability to multiply numbers by 10 and to add them up. It is also based on the understanding that multiplication can be distributed over addition and subtraction, for instance $[a - b] \times c = [a \times c] - [b \times c]$, or $[(a + b) \times c] = [(ac) + (bc)]$.

Another algorithm ("Algorithm 4") was employed to solve multidigit multiplication problems when at least one of the operands consisted of a three-digit number, e.g., in 2×3-digit or 3×4-digit multiplication. Algorithm 4 consisted of identifying one of the operands, e.g., 101 in the operation 101×965, and of multiplying it first by the units of 965 through subsequent additions, i.e., $101 \times 5 = 101 + 101 + 101 + 101 + 101 = 505$ (A). Subsequently, 101 is multiplied by the decade of 965, i.e., $101 \times 60 = 101 \times 10 = 1010$; $1010 + 1010 + 1010 + 1010 + 1010 + 1010 = 6060$ (B), and then by the hundred i.e., $101 \times 900 = 101 \times 9 = 101 + 101 + 101 + 101 + 101 + 101 + 101 + 101 + 101 = 909$; $909 \times 100 = 90900$ (C). Finally, (A), (B), and (C) are added up, $505 + 6060 + 90900 = 97465$. Algorithm 4 consists of decomposing the operations with big operands into a series of operations with smaller operands. This crucially depends on the understanding that a multiplication is equivalent to repeated addition, and on how the place notation works in multiplication. IH applied this decomposition process systematically by using a precise set of steps. For instance, one of the operands was always multiplied by the unit of the other operand first, then by the decade, and finally by the hundred. Figure 6 shows an extraordinary example of the successful use of Algorithm 4.

Figure 2 shows the proportion of use of the algorithms and of the errors they gave rise to. Although all multidigit multiplication problems were solved by using Algorithms 3, 3a, or 4, the proportion of errors was not equal across different procedures, $\chi^2(2) = 36.6, p < .001$.

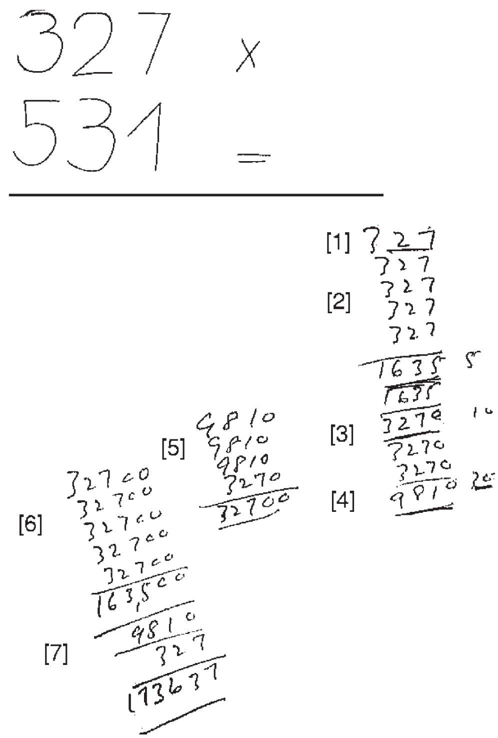

Figure 6. *An example of IH's performance in solving a multidigit multiplication operation using Algorithm 4. The operand 531 has been decomposed into the subparts 1, 30, and 500. Numbers in squared brackets indicate the main steps in performing the operation. [1] is 327 × 1. Steps [2] to [****4****] show how 327 × 30 was obtained. In particular, step [2] is 327 × 5, as indicated also by number 5 written by IH on the right-hand side to remind himself how many times he multiplied 327; [3] is 327 × 10 and [4] is 327 × 30; [5] is 327 × 100 obtained through multiple additions, [6] is 327 × 500, again obtained through multiple additions. Finally, [7] is (327 × 500) + (327 × 30) + (327 × 1). The final result is correct.*

A classification of errors is reported in Table 4. Sometimes more than one error was made in performing an operation. Errors mainly consisted of partial or impaired application of procedures and occurred also in performing multidigit division problems (34%).

IH still performed relatively well on multidigit operations of the Graded Difficulty Arithmetic Test (Jackson & Warrington, 1986).

Time-period 3: March 1999–March 2000. At time-period 3, IH's performance with multidigit addition and subtraction problems was not different from previous examinations, as those operations were performed with only a few errors. A few more mistakes were made on division problems.[3]

Conversely, results on multidigit multiplication problems indicated a significant deterioration compared to previous testing, McNemar Test, $\chi^2(1) = 2.25$, $p < .20$. In particular, most of the errors resulted from the use of long algorithms, and a new algorithm (Algorithm 5), which is a variant of a previous one, was also used at this stage. Table 4 shows the type and the proportion of errors made at time-period 3 in solving multidigit multiplication problems. The most common mistake consisted of missing part of the procedure while solving the operations (14 out of 30 errors, 47%). That is, IH thought the operation was finished and that the result produced was the final one, although it was not. This is understandable considering that the algorithms used were long and difficult to monitor.

Algorithm 5 represented a slightly longer version of Algorithm 3 and was used to solve 2×2-digit multiplication problems. It consisted of fractionating some of the steps of Algorithm 3 into further substeps. Considering again the operation 23×35 as an example, it will be remembered that at time-period 2 it was performed as follows: (1) $35 \times 10 = 350$; (2) $350 + 350 = 700$; (3) $35 \times 3 = 105$; (4) $700 + 105 = 805$.

At time-period 3, step (1) was divided into two substeps: (1a) $35 \times 5 = 175$; (1b) $175 + 175 = 350$; or, in other cases, into further substeps. For instance: (1a) $35 \times 2 = 70$; (1b) $70 \times 2 = 140$; (1c) $140 + 140 = 280$; (1d) $280 + 70 = 350$.

Similarly, at time-period 2, 48×5 used to be: (1) $48 \times 10 = 480$; (2) $480 \div 2 = 240$.

At time-period 3 the same operation was solved as: (1) $48 \times 2 = 96$; (2) $96 + 96 = 192$; (3) $192 + 48 = 240$.

Algorithm 5 is based on the knowledge of the 2 times table, on the ability to add numbers and to multiply them by 10. It is based on the distributive law, which states that multiplication distributes over addition, for instance $[(3 + 4) \times 2] = [(3 \times 2) + (4 \times 2)]$. Algorithm 5 comprises a variable number of steps, according to the complexity of the problem. The starting point always consisted of multiplying one of the operands by two and of adding up the result as many times as indicated by the other operand or by subparts of it. IH developed and applied Algorithm 5 spontaneously, clearly indicating intact knowledge of the principles pertaining arithmetical operations (i.e., arithmetical conceptual knowledge). Algorithm 5 somehow resembles what has been reported in another patient (IE; Sokol et al., 1989).

Figure 7 illustrates an example of the application of Algorithm 5. It was clearly longer than Algorithm 3 and gave rise to a significantly higher proportion of errors, $\chi^2(1) = 5.84$, $p < .02$. Figure 8 shows the proportion of use of the different algorithms in multidigit operations at time-period 3 and the number of errors resulting from each of them.

As IH used progressively longer algorithms instead of short ones, the proportion of errors increased significantly from time-period 2 to 3, McNemar Test, $\chi^2(1) = 2.25$, $p < .20$. Algorithm 3a, which had been a short-cut of Algorithm 3, was no longer used. This seems to suggest that

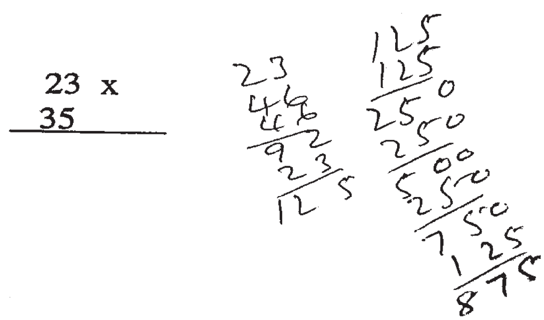

Figure 7. *An example of IH's performance in solving a multidigit multiplication problem using Algorithm 5. Note the carrying error made in the first part of his solution that is then perpetuated in the second part.*

[3] Note that performance in division problems at time-period 3 seemed to be slightly better than at time-period 2. This can be explained in terms of the reduced number of omissions in IH's answers at time-period 3.

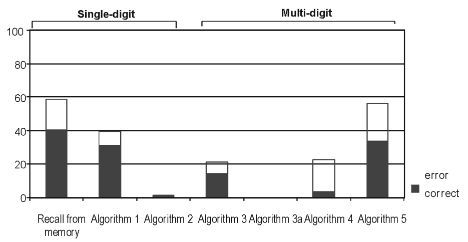

Figure 8. *Single-digit multiplication operations recalled from memory or by using Algorithms 1 and 2 (per cent of use), and multidigit multiplication operations*[a] *solved with algorithms 3, 3a, and 4 at time-period 3. The dashed areas at the top of each column indicate the proportion of errors made within each strategy used.*

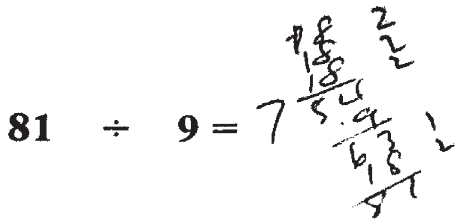

Figure 9. *An example of IH's performance in solving a division problem using Algorithm 1. The patient multiplied the dividing number (i.e., 9) by 2 (i.e., 18), and added it as many times as required to reach 81. The result (incorrect) consisted of the sum of the added numbers (i.e., 7). Note that although this algorithm was usually employed for multiplication problems, IH used it for divisions by transforming them in the reverse of multiplications. The error lies in the simple task of adding the rightmost column of 2s and 1s.*

numerical conceptual knowledge was still well preserved in IH, although his difficulties in dealing with large numbers limited the use of short algorithms. Interestingly, IH applied to division operations some algorithms that he used for multiplications. For instance, he employed Algorithm 1 to solve the operation 81 ÷ 9 (see Figure 9).

IH was not tested on the Graded Difficulty Arithmetic Test (Jackson & Warrington, 1986) at this time.

Time-period 4: August 2000. At time-period 4 of the experimental investigation, IH's cognitive decline and severe comprehension difficulties did not allow us to examine his calculation skills. The patient could only be tested on part of the Graded Difficulty Arithmetic Test (Jackson & Warrington, 1986), which showed some residual ability to perform addition problems (32% correct answers).

Conclusions on IH's performance on multidigit operations. IH's performance during approximately 3 years of investigation showed progressive decline in the ability to perform multidigit operations. This decline did not affect arithmetical operations equally: Addition and subtraction problems were relatively preserved, as were all single-digit operations, whereas IH's performance in multiplication and division operations had significantly declined over time. Despite the progressive increase of errors, the development and the use of compensatory procedures strongly suggests that IH could still well understand the principles pertaining to arithmetical operations. These algorithms seemed to compensate the loss of some memorised arithmetical times tables.

SUMMARY

This study described a 3-year follow-up (1997–2000) investigating the decline of numerical skills in a patient with semantic dementia. IH initially showed almost completely preserved numerical abilities in the context of severe disorders of semantic memory. The only impaired number skills were those highly dependent on comprehension or verbal production (e.g., defining arithmetical operations or answering number questions).

IH's performance in nonverbal number tasks was good and remained unchanged until near the end of the investigation, when severe comprehension problems possibly interfered with the patient's ability to perform number tasks. Similarly, he performed quite well on verbal number tasks for the first 2 years of the investigation. Towards the end of the examination, IH's performance declined dramatically in many tasks (e.g., counting), some transcoding tasks could no longer be performed (e.g., number words from numerals), and he made several mistakes with long or complex numbers. Nevertheless, IH could still compose the value of numbers with tokens (up to two-digit), and perform simple arithmetical operations.

DISCUSSION

Two major results emerged from the 3-year follow-up study of IH's number abilities: First, arithmetic conceptual knowledge was well preserved despite severe impairment of nonarithmetic conceptual knowledge. Second, the patient's progressive decline revealed patterns of dissociations between different number abilities.

Selective preservation of conceptual knowledge of arithmetic

IH repeatedly demonstrated good understanding of arithmetical operations and of the principles pertaining to them, which sharply contrasted with severe impairment on all the other types of conceptual knowledge. This distinction is quite remarkable given the severity of his semantic disorder affecting the simplest of tasks, and considering the complexity of arithmetical conceptual knowledge. Although the use of compensatory strategies has been reported in other patients, IH is the first patient with a reported dissociation *between* different types of conceptual knowledge, as arithmetical conceptual knowledge has always been previously reported in the context of preserved general semantics (Delazer & Benke, 1997; Hittmair-Delazer et al., 1994; McCloskey et al., 1985; Sokol & McCloskey, 1990, 1991; Sokol et al., 1989; Warrington, 1982). A subsequent study to our original report (Cappelletti et al., 2001) also reported a patient with semantic impairment and preserved number skills, although arithmetical conceptual knowledge was not the focus of this investigation (Crutch & Warrington, 2002).

As we said in the Introduction, conceptual knowledge seems to require a higher level of comprehension of numerical concepts as compared to other numerical competences such as counting or reading numbers aloud. This study showed that despite cognitive decline, the core number semantic abilities remained intact in IH, as demonstrated by his adaptable mastery of arithmetical principles and his ability to manipulate quantities. IH used relatively fewer and longer algorithms in multidigit multiplications compared with other patients (for example, patient BE, Hittmair-Delazer et al., 1994). This seems to suggest that IH's arithmetical conceptual knowledge was *relatively* well preserved compared to other type of non-numerical knowledge, and it was indeed less good than that of other patients. It is also important to remember that IH's degenerative disorder might have reduced the cognitive resources that are needed to process several alternative strategies.

It may be tempting to suggest that IH's numerical skills could be explained in terms of his pre-morbid experience with numbers. It will be remembered that IH used to be a banker and enjoyed for many years other number-based activities such as gambling and lottery. These activities imply familiarity with numbers, and occasionally they require simple calculation with multidigit multiplication problems. However, many other jobs, like shopkeeping, accountanting, and even the university teaching, may require more practice in performing arithmetical operations. Although it may be possible that pre-morbid experience with numbers helped IH to maintain his number skills, we repeatedly found that patients with neurodegenerative disorders and with no specific pre-morbid experience with numbers showed significantly better preserved numerical abilities compared to other cognitive skills and to other semantic domains (Cappelletti, 2002; Cappelletti, Butterworth, & Kopelman, 2004a). Moreover, it does not seem very plausible to explain IH's use of complex algorithms as resulting from his pre-morbid experience with numbers. For instance, when initially tested, the patient was well able to perform arithmetical operations, including multidigit multiplication problems, by using canonical procedures. Hence, IH knew how to perform arithmetical operations with a canonical procedure, and the complex algorithms he employed later were exclusively due to his inability to use simpler and more efficient procedures.

We have attempted to explain the dissociation between arithmetical and nonarithmetical knowledge in terms of the neuroanatomical substrates that seem to be engaged in processing numerical and nonnumerical information in normal subjects (Cappelletti, Kopelman, & Butterworth, 2002). These can be broadly identified in the areas around the parietal lobes and the left inferior temporal lobe, respectively (Dehaene, Piazza, Pinel, & Cohen, 2003; Martin, Wiggs, Underleider, & Haxby, 1996; Mummery, Patterson, Hodges, & Price, 1998). Hence, the understanding of arithmetical concepts might be preserved when the parietal areas are intact, as in patient IH. It should be noted, however, that impaired arithmetical conceptual knowledge has been associated with both intact and lesioned parietal areas in neuropsychological patients (Delazer & Benke, 1997; Hittmair-Delazer et al., 1994). Additional neuropsychological and neuroimaging investigations are needed to explore this issue further.

Progressive decline revealed dissociations between number abilities

One type of dissociation that emerged in IH's performance is between multiplication and other arithmetical operations. IH made most of the errors in multiplication problems, and this became progressively worse through time. It might be suggested that IH's impairment with multiplication problems consists of an artefact of task difficulty. Indeed, even normal subjects find multiplication problems difficult, and deficits in performing multiplication problems have been often reported in neuropsychological patients (for example, Dehaene & Cohen, 1997; Grafman et al., 1989; McCloskey et al., 1991), although selective preservation for multiplication facts is also on record (Delazer & Benke, 1997; Van Harskamp & Cipolotti, 2001). For at least two reasons the issue of task difficulty does not seem to account for IH's impairment with multiplication facts. First, it may be remembered that the patient was indeed good at retrieving some of the times tables: many of the algorithms IH used were based at least on the $N \times 2$ and $N \times 10$ tables. Therefore, he seemed to have lost some of the multiplication facts, in particular those involving large operands, but had good knowledge of at least some others. Second, the algorithms that IH spontaneously devised and used were far more complex and articulated than the canonical procedures that he could no longer use.

At an anatomical level, the dissociation between multiplication and other arithmetical operations in patient IH raises interesting issues for Dehaene and Cohen's model (1995). This assumes that the retrieval of multiplication tables relies on the inferior parietal lobule (SMG/AG in particular) and on a subset of left-hemispheric language areas. IH's impaired arithmetical facts seems compatible with the hypothesis that these facts rely on verbal memory, as the patient's language abilities were severely impaired, but not with Dehaene and Cohen's prediction (1995) that the left inferior parietal lobule plays a critical role in the retrieval of multiplication facts. IH's inferior parietal areas were well preserved despite impaired multiplication facts, suggesting that the temporal areas may play a more crucial role than initially hypothesised by Dehaene et al. (2003; Dehaene & Cohen 1995).

The second type of dissociation that emerged in IH's performance was between impaired arithmetical procedures and simple facts on the one hand, and preserved conceptual knowledge of arithmetic on the other. The impaired use of arithmetical procedures and simple facts "indirectly" revealed intact understanding of the meaning of arithmetical operations in IH. Briefly we can note the use of the commutative law in the latter phase of performance (2×9 and 9×2 were both solved as $9 + 9$); the use of the distributive law is illustrated in Figure 7; $23 \times 35 = (23 \times 5) + (23 \times 30)$.

A more explicit assessment of arithmetical conceptual knowledge, as has been done with other patients (e.g., Delazer & Benke, 1997), might have been impossible in patient IH. For example, problems such as: "If $12 \times 4 = 48$, $48 \div 4 = ?$" or "If $12 \times 4 = 48$, $120 \times 40 = ?$" (Delazer & Benke, 1997) require a much higher level of comprehension that made it impossible to test them in IH (for instance, the results of the second operation does not have to be calculated but rather is derived from the first). Although dissociation between arithmetic conceptual knowledge and other numerical abilities have been previously reported in other patients (Delazer & Benke, 1997; Hittmair-Delazer et al., 1994), it has never been described in terms of impaired nonarithmetic conceptual knowledge, and in the context of a progressive cognitive disorder. More importantly, IH's performance showed not just his excellent understanding of arithmetical operations, but also the ability to adapt the algorithms to contingent impairments, showing flexible and skilled mastery of arithmetical conceptual knowledge. For instance, he knew that $(N \times 10) = (2N \times 5)$, that $(N \times M) = (N/2 \times 2M)$, and that $(N \times M) = (N + N + N + \ldots$ up to the number of Ns denoted by M).

Third, a dissociation emerged within transcoding abilities. IH's transcoding skills declined slowly over the 3 years of the study. Initially, five- and six-digit numbers were the only ones impaired, but towards the end of the investigation only single digits could be reasonably well performed. Difficulties in processing long numerals (in terms of number of digits) have been reported in neurological patients (e.g., Macoir et al., 1999; Noel & Seron, 1995), and occur in the normal development of transcoding rules as they follow the length of the numerals (Noel & Turconi, 1999). Long multidigit numbers consist of a quite complex lexico-syntactic structure based on the aggregation of *sum* (e.g., "three hundred and two" means "three hundred plus two") and *product* (e.g., "three hundred" means "three" times "one hundred") relationships within numbers. These complex stimuli are more error prone than simpler ones, and they were therefore more vulnerable to IH's cognitive decline.

Towards the end of the investigation, IH was also quite impaired at writing number words, which have been well preserved for a long time (Butterworth, Cappelletti, & Kopelman, 2001; Cappelletti et al., 2002). The incapacity to write numbers in verbal format extended to all items, with the exception of the word "hundred." Impaired writing of number words cannot be explained in terms of sensory-motor impairments or difficulties in remembering number strings, as they would have equally affected IH's performance with Arabic numbers. It may be possible to think that writing number words is more difficult than Arabic numbers, although this does not explain why IH could write the number word "hundred," which is quite long and relatively less frequent than other number words such as "one" or "six." Alternatively, we might think that IH's impairment was in shifting from an Arabic to a verbal format, similarly to other reported patients (Della Sala, Gentileschi, Gray, & Spinnler, 2000; Thioux, Ivanoiu, Turconi, & Seron, 1999). We do not, however, have any independent evidence (i.e., in a non-number task) indicating that a generalised shifting problem occurred in IH.

To sum up, we have reported a 3-year follow-up study describing the progressive deterioration of number knowledge in a semantic dementia patient. Two major results emerged: First, the patient's numerical conceptual knowledge was well preserved despite severe impairment of non-numerical conceptual knowledge. Second, IH's progressive decline revealed interesting patterns of dissociations between different number abilities. These were between multiplication and other arithmetical operations, between impaired knowledge of number facts and procedures on one hand, and conceptual knowledge of arithmetical principle on the other, and between different transcoding skills. A few other studies on the decline of number skills have been reported in other patients with progressive disorders, and dissociations between different types of number abilities have been described in nonprogressive disorders. This study has suggested how progressive cognitive decline can be used in neuropsychology to reveal patterns of dissociations among cognitive skills.

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APPENDIX A

Nonverbal number knowledge

Dot tasks

1. *Dot seriation*. IH was required to order three cards each with a different number of dots from the smallest to the biggest (e.g., fromx • • • •, • •, • • • to • •, • • •, • • • •). Three different sets of six triplets each ($N = 18$) were randomly presented to the patients: in the first six, each card contained 1 to 5 dots, in the second 6 to 10 dots, and in the third 1 to 10 dots.
2. *Mangnitude comparison*. IH was asked to compare the magnitude of two sets of black dots randomly arranged on pairs of cards (e.g., • • • vs • •). Eighteen pairs were organised in three different sets: in the first, each item of the pair consisted of 5 or fewer dots, in the second 6 to 10 dots, and in the last set 1 to 10 dots. No explicit knowledge of numbers words was required for this task, although the understanding of magnitude was needed.

Number seriation

Eighteen triplets of single-digit numerals were randomly presented in large font, each numeral in the centre of a card. IH was asked to reorder the items in each triplet from the smallest to the biggest according to their magnitude (e.g., from 6, 9, 3 to 3, 6, 9). Three different sets of triplets were used: the first six comprised numbers between 1 and 4; the second, numbers between 5 and 9, and the third numbers between 1 and 9.

Number comprehension

1. *Magnitude comparison*. Twenty pairs of one- to four-digit numerals (e.g., 3098 vs. 3089) were presented to the IH, who was asked to point or to name the larger numeral in each pair. Pairs of numerals were printed in font 24 in lower-case on separate cards and each pair presented together for comparison.

2. *Number composition with tokens*. Forty-eight two- and three-digit numbers were orally presented by the examiner. IH was asked to select the tokens to the value of each number. Tokens consisted of plastic coloured rounded chips with numerals ranging from 1 to 500 written on them. Tokens were left in front of IH in random order. After describing each token and naming its value, the experimenter said a number aloud and showed IH how to compose its value by choosing the correct tokens. If IH understood the instructions, the experimenter started the task. There was no time restriction to produce an answer.
3. *Placing numbers on an analogue number line*. The patient was presented with 36 identical black lines each with the indication of the extremes. Three different types of extreme were used, 0–10, 0–100, or 0–1000. For each of the 36 lines, IH was presented with a numeral and required to indicate its *approximate* position along the line using the extremes as a reference point.

Verbal number knowledge

Number tasks

Sequence knowledge

1. *Dot enumeration*. IH was presented with 10 cards with patterns of dots ranging from 1 to 10 on each of them (e.g., • • • • or • •). Each card was presented twice in a random fashion ($N = 20$). IH was asked to say aloud the number of the dots on each card. There was no time limit to the stimuli presentation.
2. *Counting*. IH was asked to count forward by one from 1 to 20, by one from 111 to 120, by 2 from 21 to 39, and backward from 20 to 1 ($N = 60$).
3. *"What comes next?", "What comes before?"*. Forty single- to four-digit spoken numbers were randomly presented to IH, who were asked to produce the number coming before or after each of them (e.g., "What comes after 899?").

Number recognition

These tasks aimed at excluding the existence of difficulties in recognising numbers. The stimuli consisted of 3 single-digit numerals (e.g., 5), 3 numbers between 11 and 19 (e.g., 17), three decades (e.g., 40), 3 two-digit numbers between 21 and 99, excluding the decades (e.g., 34), 3 three-digit (e.g., 809), and 3 four-digit numbers (e.g., 6720). IH was tested using two conditions.

1. *Matching numerals to spoken numbers*. IH was asked to match 18 spoken numbers to the corresponding numerals presented together with three alternatives. The first was a number of the same category of the target one (i.e., they were both units, or two-digit numbers); the other two were numbers of different categories. For example, number 2 was presented for the target 5; 50 and 500 for the target 5.
2. *Matching written number names to spoken numbers*. IH was asked to match 18 spoken number words (e.g., "seven") to the corresponding written number words (e.g., seven) presented together with three alternatives of the same type of those presented in the previous task.

Number comprehension

Bisection task. IH was orally presented with 10 pairs of single- to four-digit numbers and asked to provide the number between them ($N = 10$). For example, he was required to say 206 for the pairs 205 and 207. In addition, a non-numerical version of the task was used, which differed from the previous one only in terms of the material used. The patient was presented with pairs of letters of the alphabet, of months of the year, and of days of the week, and asked to say the middle one ($N = 30$). The task aimed at examining whether IH could equally perform the tasks with numerical and non-numerical items.

Transcoding

Six transcoding tasks from written or spoken number words to numerals and vice versa were administered to the patient. A set of 100 items was used. The stimuli consisted of 10 single-digit numbers (0 to 9), 10 numbers between 10 and 19, 20 two-digit numbers between 20 and 99 (8 ending in 0, 12 not ending in 0), 30 three-digit numbers between 100 and 999 (15 ending in 0 or with internal 0, and 15 without 0), and 30 four-digit numbers between 1000 and 9999 (15 ending in 0 or with internal 0, 15 without 0). When a written presentation was required, each item was printed in font 24 in lower case (in case of number words) on separate cards and presented to the patients in random order. In spoken presentations, stimuli were presented one at the time by the experimenter.

1. *Numerals to spoken numbers* (i.e., reading "1"). Numerals were written in the centre of a card and randomly presented to the patient for reading aloud ($N = 100$). Twenty five- and six-digit numbers were also presented. There was no time limit to the stimuli presentation.

2. *Written number words to spoken numbers* (i.e., reading one). Written number words were randomly presented to the patient for reading aloud. The stimuli ($N = 50$) were selected from the set of numerals used in the previous reading task.
3. *Spoken numbers to numerals* (i.e., writing "1"). Some of the numerals used for the reading task were presented to the patient for writing to dictation ($N = 50$). Each stimulus was dictated aloud by the experimenter for a written response.
4. *Spoken numbers to written number words* (i.e., writing "one"). Twenty number words were selected from the set of numerals used for the reading task. The new set included an equal number of items from each type of stimulus (i.e., single, teens, tens, etc). Stimuli were dictated one at a time by the experimenter and IH was asked to write them in verbal format.
5. *Numerals to written number words* (i.e., 1 → one). IH was presented with 35 numerals, one at a time, and asked to write the number words corresponding to each of them.
6. *Written number words to numerals* (i.e., one → 1). The reverse task was administered to the patients. They were presented with 35 written number words corresponding to the set previously used, and asked to read them and to write the correspondent numerals.

Special transcoding tasks

A set of transcoding tasks was especially designed to investigate whether the presence of zeros in multidigit numbers might increase the proportion of errors. Numbers ending or containing zeros were part of the experimental set used before, although they were presented in combination with single-digit numbers or other multidigit numbers with no zeros. The new set of items contained only three- and four-digit numbers including or ending in zero ($N = 20$). IH was administered three tasks.

1. *Spoken numbers to numerals* (i.e., writing 302). Each stimulus was dictated aloud by the experimenter for a written response ($N = 20$).
2. *Written number words to spoken numbers* (i.e., reading aloud "three hundred and two"). The set of written number words were randomly presented to IH for reading aloud ($N = 20$). There was no time limitation to the presentation of the stimuli.
3. *Written number words to numerals* (i.e., three hundred and two → 302). IH was presented with the 20 written number words used in the previous set, and asked to write the numerals corresponding to each of them.

Encyclopaedic knowledge

1. *Personal number facts*. IH was presented with 10 questions tapping personal numerical information (e.g., "How old are you?"), and asked to produce a spoken answer.
2. *Nonpersonal number facts*. IH was presented with 10 questions tapping nonpersonal numerical information (e.g., "How many months are there in a year?") and asked to produce a spoken answer.
3. *Naming and writing arithmetical signs*. The four arithmetical signs (i.e., $+ - \times \div$) were presented in written and spoken formats and IH was asked to name and write them.
4. *Definition of arithmetical operations*. The patient was asked to define the four arithmetical operations (e.g., "Could you tell me what is an addition?").

Calculation tasks

Arithmetical facts

Three hundred single-digit arithmetical operations (e.g., $2 + 1$ or 3×2) were orally presented to IH and an oral answer was required. IH was first administered all the addition problems from $0 + 0$ to $9 + 9$ ($N = 100$), followed by the multiplication problems ($N = 100$). In these tasks, small numbers were equally presented in the first or in the second position. A different but similar set was used for a subtraction ($N = 54$) and a division task ($N = 50$).

Mental calculation

The Graded Difficulty Arithmetic Test (Jackson & Warrington, 1986) was used, consisting of a set of two- and three-digit addition and subtraction problems ($N = 28$), partially requiring carrying and borrowing (e.g., $15 + 13$ or $128 + 149$). According to the task's instructions, a mark should be assigned if a correct answer is produced within 10 seconds. As the patient was generally very slow at performing cognitive tasks, he was not always timed while doing this task.

Written calculation

Written arithmetical operations were presented to the patient, who was asked to produce a written answer. Groups of four operations were written on an A4 paper. Once solved, each operation was covered by the experimenter to avoid any influence on the others.

1. *Addition problems*. Thirty-two problems with addends consisting of two- and three-digit numerals were presented to IH, half requiring carrying (e.g., $624 + 277$).
2. *Subtraction problems*. Thirty-two problems with two- and three-digit numerals were administered to IH, half requiring borrowing (e.g., 742–393).

3. *Multiplication problems*. Eighteen problems with two and three-digit numerals were presented (e.g., 468 × 35).
4. *Division problems*. Twenty problems with single- and two-digit numerals were administered (e.g., 36 ÷ 12).

Special calculation tasks
An additional calculation task was used to explore the use of arithmetical rules in more details. Twenty single-digit arithmetical problems were orally presented by the examiner, and IH was asked to produce a spoken answer. Problems were all based on rules, such as adding or subtracting 0 (e.g., 6–0 or 5 + 0), and multiplying numbers by 0 or 1 (e.g., 7 × 0 or 2 × 1).

Approximate calculation
Twenty-four arithmetical problems consisting of two- and three-digit numerals were presented along with four different results. The first was very close to the correct one, the second was approximately double the correct result, the third was very far from the correct result, and the last consisted of the result of an operation with the given operands but a different arithmetical sign. For example, the operation 258 + 144 was presented with the following options: 400, 800, 58, 114. For each arithmetical operation, IH was asked to decide as fast as possible the option that was *approximately* the closest to the correct result. A few examples—different from the experimental set—were given to IH to clarify the instructions and to let him familiarise with the procedure.