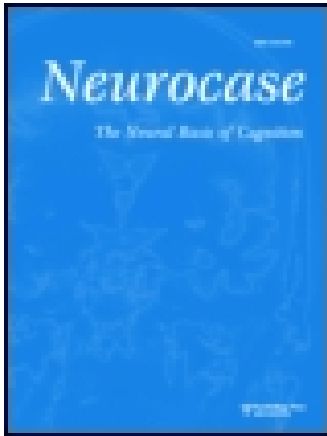


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Pure Global Acalculia Following a Left Subangular Lesion

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Abstract

We describe the case of a right-handed patient who presented a severe acalculia in the context of a pure Gerstmann syndrome following a subangular lesion that spared the left inferior parietal lobule (IPL). The patient showed impairments in Arabic and verbal codes, in number production and comprehension, as well as in numerical facts and problem solving. By using the EC301 calculation battery, semantic and syntactic tasks in Arabic and verbal codes, we tested the different hypotheses raised by the cognitive neuropsychological models of acalculia. The patients' difficulties, which were not associated with a general intellectual deterioration, and those affecting number processing as a particular semantic class, were indicative of a "global acalculia". This deficit, which exceeded the anarithmetia usually described in Gerstmann syndrome following left IPL lesion, suggested that the isolation of this area may constitute a sufficient condition for producing such a global acalculia. These results are discussed in terms of a disorder in the manipulation of mental images of spatially related objects.

Introduction

The term acalculia was introduced by Salomon Henschen in 1920 (Henschen, 1920) to designate an acquired impairment in calculation and number processing in the absence of other cognitive disorders. A few years later, Joseph Gerstmann (Gerstmann, 1924, 1927, 1930) described a new syndrome composed of agraphia, left-right disorientation, finger agnosia and acalculia which resulted from a left parietal lesion. Subsequently, Hécaen and colleagues (Hécaen *et al.*, 1961) identified three types of calculation deficits: *visuo-spatial acalculia* (referring to an erroneous alignment of figures in space during the calculation), *anarithmetia* (referring to the loss of the capacity to solve operations and to deal with quantities) and finally *aphasic acalculia* (resulting from a language impairment). Since then, the dissociation between language and numerical processing has been observed a number of times in aphasic patients whose counting abilities were preserved (for a review see Thioux *et al.*, 1998), as well as in a case of semantic dementia (Cappelletti *et al.*, 2001). The reverse pattern of a selective impairment in numerical processing without language impairment has also been reported although less frequently (Cipolotti *et al.*, 1991; Takayama *et al.*, 1994). There is therefore a double dissociation between language and number processing implying different structures for these functions. For example, in patients with left parietal lesions, isolated acalculia was attributed to a deficit in working memory processes (Takayama *et al.*, 1994). In another case report, selective

deficits in the production and comprehension of numerals beyond the number 4 (despite the preserved understanding of physical quantities) were interpreted as a very circumscribed category-specific impairment in the semantic system (Cipolotti *et al.*, 1991). Numbers are thus considered as a particular semantic class serving three different meanings: i) a *numerical meaning* concerning measure, cardinality and ordinality, ii) a *sequential meaning* used in counting and reciting verbal series such as days of the week, name of the month or number of the month and iii) a *non-numerical meaning* referring to memorised numbers that denote, for example, to fixed anniversary dates (i.e. July 4th or September 11th), or TV channel number (Fuson, 1988; Hittmair-Delazer *et al.*, 1995).

Cognitive neuropsychology has proposed different theoretical models for number processing and calculation in which Arabic and verbal (oral and printed word) numbers are separated into different domains, each according to their own rules (Deloche and Seron, 1987; MacCloskey and Caramazza, 1987; Dehaene, 1992; Dehaene and Cohen, 1995). The model of MacCloskey and Caramazza proposes that number representation is abstract and amodal and defines two separate systems for number processing and calculation (MacCloskey and Caramazza, 1987). In the *number processing system*, comprehension (input) and production (output) form two distinct modules sharing the same cognitive architecture. Inside these modules, verbal and Arabic numbers are separated. The category of oral verbal numbers is

characterised by lexical, phonological (or graphemic for printed word numbers) and syntactic components. The syntactic component, which is also present in Arabic numerals, refers to the processing of relations among the digits in order to produce or to understand a number as a whole. The *calculation system* deals with long-term memory storage of numerical facts (counting, multiplication tables, etc...), knowledge of arithmetical symbols and problem solving procedures. Together, the number processing (comprehension and production) and calculation systems are linked by an abstract amodal semantic representation.

In the “triple code model” of Dehaene, the notion of an amodal and abstract internal representation for both number processing and calculation was questioned (Dehaene, 1992; Dehaene and Cohen, 1995). This model, revised on the basis of several observations (Dehaene and Cohen, 1997; Cohen *et al.*, 2000), distinguishes three intimately linked codes. The *Arabic visual code*, where numbers are represented by series of digit succession, depends on the right and left occipital lobes. The *auditory-verbal code*, linked to phonologic and graphemic forms, is localised in the left temporal lobe and in left subcortical structures and supports rote-learned arithmetical facts and counting. Finally, the *analogical magnitude code* localised in bilateral parietal areas, involving the inferior parietal lobule, supports the semantic information such as quantity comparisons. These authors initially based their model upon the case of a severe aphasic patient (NAU) who failed on arithmetic facts and also on exact calculation, but remained capable of making approximations (Dehaene and Cohen, 1991). From this case, the authors assumed that the patient’s approximation capabilities were processed by his preserved right hemisphere. Later, in an experimental study, the authors stressed the role of the bilateral inferior parietal lobules in approximations (Stanescu-Cosson *et al.*, 2000). Moreover, they presented two left hemisphere-damaged patients, BOO (Dehaene and Cohen, 1997) and ATH (Cohen *et al.*, 2000) who both suffered from aphasia in association with an impairment in the processing of arithmetical facts and verbal numerics. In contrast, MAR (Dehaene and Cohen, 1997), a left-handed patient who suffered from a right parietal lobule lesion, was impaired only in the manipulation of quantities although his performance on arithmetic facts and verbal numerical tasks was normal. Based on these observations, Cohen *et al.* (Cohen *et al.*, 2000) distinguished two types of anarithmetia: a verbal one and a semantic one. The “*verbal anarithmetia*” is associated with aphasia including damage to cortical areas and cortico-subcortical loops. The “*semantic anarithmetia*” without aphasia occurs in the context of a Gerstmann syndrome (GS) following an inferior parietal lobule lesion.

We have recently reported the case of a patient (HP) who presented GS following a subangular lesion (Mayer *et al.*, 1999). This case was considered particularly pure in comparison to other published cases. Furthermore, his acalculia appeared to be quite specific. With respect to the other constituting symptoms of GS, the acalculia observed in this patient was disproportionate and global. Indeed, despite the

complete absence of other intellectual and language deficits, the impairments severely affected all aspects of calculation and number processing in both Arabic and verbal codes (i.e. number production and comprehension, as well as numerical facts and problem solving). This global number processing deficit appeared particularly suited to an in-depth investigation of the different hypotheses and cognitive neuropsychological models of acalculia. We first tested the patient’s semantic memory to verify whether or not his acalculia represented a category-specific semantic deficit as previously suggested (Cipolotti *et al.*, 1991). We then sought to verify whether HP’s performances in transcoding tasks in both verbal and Arabic codes relied on an amodal semantic representation (MacCloskey and Caramazza, 1987). Finally, we tried to determine how such an extensive acalculia could fit with the “triple code model” predictions in GS (Dehaene, 1992; Dehaene and Cohen, 1995).

Case history

This patient’s case history has been described in detail in a previous report (Mayer *et al.*, 1999), and will only be summarised briefly here. HP was a 59 year-old, right-handed man who worked as an insurance agent until his stroke in 1994. He had completed secondary school education and had completed one year in university. Before the stroke, he never presented difficulties with numbers or letters (spelling) and was used to dealing with contracts involving figures and calculation. He was admitted to Geneva University Hospital following the abrupt onset of writing and calculation disturbances and difficulties in dialing phone numbers. Neuropsychological testing revealed the presence of all four symptoms of GS, i.e. agraphia, acalculia, finger agnosia and right-left disorientation. MRI evaluation revealed a small subangular focal subcortical white matter lesion undercutting the left angular gyrus (Fig. 1). HP’s lesion, which clearly spared the inferior parietal lobule (IPL), isolated the anterior part of the angular gyrus from temporo-occipital and other parietal areas. Finally, it should be noted that the lesion did not extend to the thalamus.

Neuropsychological evaluation

The psychometric data, obtained three months following admission, can be found in Mayer *et al.* (1999). Here we have selected the language tests that are most relevant for testing the cognitive hypotheses. HP was oriented, his spontaneous speech was fluent and he had no aphasia as confirmed by his scores (see Table 1) obtained on the French version of the Boston Diagnostic Aphasia Examination (BDAE) (Goodglass and Kaplan, 1972; Mazaux and Orgogozo, 1982). However, HP had mild difficulties in reciting rote-learned lexicons such as days of the week or months of the year. He produced them without errors, but slowly and hesitantly. Specific difficulties in spelling and writing to dictation were characterised by substitutions only between lower case letters b/p, q/d. HP showed a peripheral dysgraphia

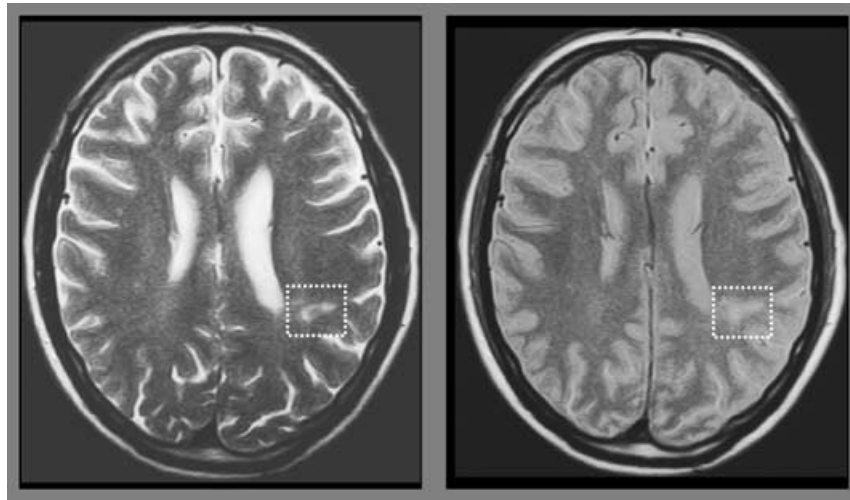


Fig. 1. Illustration of the subangular lesion that spared the left inferior parietal lobe in patient HP. Neuroradiological evaluation was done with MRI T2 (left) together with T1 (right) images shown here on axial slices for the best demonstration of the lesion site.

Table 1. Language examination: BDAE. Specific difficulties in spelling and writing to dictation were due to substitutions between lower case letters b/p, q/d

Sub-tests	Scores
Fluency:	
Articulation rating	7/7
Phrase length	7/7
Verbal agility	14/14
Automatic speech:	
Automated sequences	7/9
Reciting	2/2
Repetition:	
Words	10/10
High probability sentences	8/8
Low probability sentences	7/8
Naming:	
Responsive naming	30/30
Picture naming	100/105
Body parts	18/20
Semantic fluency	23/23
Auditory Comprehension:	
Word discrimination	72/72
Commands	13/15
Body parts	19/20
Complex ideational material	11/12
Reading and comprehension:	
Words	30/30
Sentences	9/10
Words-pictures matching	10/10
Oral spelling	7/8
Sentences and paragraphs	10/10
Writing:	
Serial writing	45/47
Spelling to dictation	6/10
Sentences to dictation	9/12
Narration	3/4

with an impairment of his motor graphic patterns (Martory, 1996; Zesiger *et al.*, 1997)

Concerning the other most relevant neuropsychological scores, HP obtained a memory quotient (MQ) of 114 on

the long term memory tests of the WMS-R (Wechsler Memory Scale-Revised). His scores were also good on the Rey auditory verbal learning test and the Rey visual design test (percentile 60 for both). Short-term memory was impaired; digit span was only four forwards and two backwards but the “Brown-Peterson” Auditory Consonant Trigrams Test for the evaluation of working memory (Brown, 1958; Peterson and Peterson, 1959) was normal. On the WAIS-R (Wechsler Adult Intelligence Scale-Revised), selective impairments appeared in subtests necessitating calculation and number processing; the verbal intelligence quotient (IQ) prorated without the digit span and arithmetic subtests was good (cf. Mayer *et al.*, 1999).

In order to investigate more precisely the patient’s capacity to process semantic information, we carried out other specific tests. His performances on semantic tasks, summarised in Table 2, were all normal. Word-fluency tasks, used to evaluate the spontaneous production of words beginning with a given letter (e.g. P; phonemic fluency, $n = 24$) or a given category (e.g. animals; semantic fluency, $n = 35$) within two minutes, were normal (Cardebat *et al.*, 1990). HP was able to name 25 living objects on the basis of either a “visual” (e.g. for a ladybird: “*a red insect with black dots*”, 25/25) or a “functional” definition (e.g. for a ladybird: “*an insect that is thought to bring good luck*”, 25/25). He was also able to define a set of ten concrete and abstract words, to designate the synonym of an abstract word among a choice of four words, and to explain commonly used sayings. Finally, sentence comprehension, assessed by an abridged version of the Token Test (De Renzi and Faglioni, 1978) and the Montreal-Toulouse Test (MT86) (Nespoulous *et al.*, 1992), was almost perfect (35/36 and 40/41 respectively).

Assessment of numerical processing

Difficulties in calculation constituted HP’s main complaint. He claimed that: “*for letters, there is a consistency, for numbers, I cannot link them to anything, I do not see what it is*”. The patient had lost his multiplication tables and was

Table 2. Language examination: semantic tasks. Oral naming was first evaluated by the *Exadé test* (Bachy-Langedock, 1989) and an abridged version of the *Boston Naming Test* (Colombo-Thuillard and Assal, 1992). In the *part of object naming* task, 25 parts of pictured objects were presented one by one (e.g. a heel, a handle). A set of 48 images belonging to six different categories (clothes, tools, furniture, vegetables, fruits, animals) of living/non living objects (Snodgrass and Vanderwart, 1980), were used for naming, designation (one out of eight) and classification tasks

Sub-tests	Performance
Oral naming (N of correct):	
Exadé	90/90
Boston Naming Test	34/34
Parts of objects	25/25
Living category	24/24
Non-living category	24/24
Picture designation and classification (N of correct):	
Designation	48/48
Classification	48/48

unable to solve simple arithmetical problems; addition was possible only by counting on his fingers, the procedure for subtraction was impaired. In reading tasks using Arabic code, the patient sometimes produced number sequences that did not exist in French (e.g. for 842 he said “*huit cent quarante deux cent*/eight-hundred and forty-two hundred”).

An extensive evaluation of number processing was carried out using the EC301 protocol (Deloche and Seron, 1991, see also Dellatolas *et al.*, 2001). This battery includes thirteen tasks assessing the different ways in which quantity processing and calculation skills can be resolved and emphasises transcoding tasks. The EC301 protocol was administrated to the patient six months after his stroke. The results are detailed in Table 3 and commented below.

Oral and written counting (C1-C3). In the oral and written counting tasks, HP counted on his fingers. Oral one-by-one counting was very slow, the numbers 12 and 19 were omitted (counting from 1 to 31). Counting three by three, ten by ten, or backward was impossible (from 22 to 1). Counting in Arabic or alphabetic code was also impaired.

Dot counting (C4-C8). HP made dot-counting errors specially when there were more than five randomly distributed elements.

Transcoding tasks (C9-C15). Repetition, reading and writing to dictation in alphabetic code were perfect (12/12 in each). Reading and writing in the Arabic code were severely impaired (6/12 and 4/12). Transcoding from Arabic to alphabetic code or back was very difficult for the patient (6/12 and 2/12).

Naming and writing to dictation of arithmetic symbols (C16-C17). Both naming and dictation were hesitant but accurate.

Magnitude comparison (C18-C19). In these tasks the patient was asked to compare two numbers and to point to the greater. In Arabic code, he failed only for items presenting the same number of digits and a high degree of visual similarity (i.e. 329 and 325). When HP performed correctly,

Table 3. Calculation and number processing: EC301 battery

Sub-tests	Scores
Oral and written counting:	
C1: Spoken verbal counting	3/8
C2: Arabic digit counting	1/2
C3: Written verbal counting	1/4
Dot counting:	
C4-C8: Enumeration of dots	25/30
Transcoding tasks:	
C9: Oral repetition of numbers	58/84
C10: From Arabic to alphabetic code	12/12
C11: Reading aloud in Arabic code	10/12
C12: Writing to dictation in alphabetic code	6/12
C13: Reading aloud in alphabetic code	12/12
C14: Writing to dictation in Arabic code	12/12
C15: From alphabetic to Arabic code	4/12
Naming and writing to dictation of arithmetic symbols:	
C16: Arithmetical signs: naming	2/12
C17: Arithmetical signs: writing to dictation	8/8
Magnitude Comparison:	
C18: Magnitude comparison of Arabic numbers	6/8
C19: Magnitude comparison of alphabetic numbers	8/8
Oral and written mental calculation:	
C20: Mental calculation on verbal numbers	12/16
C21: Mental calculation on Arabic numbers	4/16
Approximation of arithmetic results:	
C22: Approximation of the result of an operation	6/16
Positioning a quantity on a scale:	
C23: Placing a number in the Arabic code on a scale	2/16
C24: Placing a number in the Verbal code on a scale	6/16
Writing an operation:	
C25: Placing multi-digit numbers to perform an operation	2/16
Written calculation:	
C26: Written calculation: additions	6/8
C27: Written calculation: subtractions	0/4
C28: Written calculation: multiplications	0/4
Perceptive estimation of quantities:	
C29: Magnitude approximation of pictures	2/7
Cognitive estimation of quantities and precise numerical knowledge:	
C30: Contextual magnitude judgments	4/12
C31: Precise numerical knowledge	10/10
	10/12

his strategy was always the same: he counted one by one the digits of each item and assumed that the larger item corresponded to the larger quantity. In the alphabetic code, the comparisons were highly impaired since HP relied on visual and lexical cues. For example, he concluded that “*cent mille soixante cinq*/a hundred-thousand and sixty-five” was larger than “*trois cent mille*/three-hundred thousand” because: i) the first number was composed of four words instead of three in the second and because ii) the first word “*cent*/one hundred” in the first item was larger than the first word “*trois*/three” in the second item.

Oral and written calculation (C20-C21). These tasks are composed of 16 basic operations (addition, subtraction, multiplication, and division) relying on long-term memory. Since HP was not capable of carrying out such operations mentally, he was allowed in many instances to perform the

task relying on his own strategies. In the present case, HP adopted the following method: first, he drew dots on a sheet in a configuration corresponding to the question (e.g. for $3 \times 4 =$ three lines of four dots), then he counted all the dots using his fingers. He was unable to recite any of the multiplication tables, even the table of two, arguing that “*he had lost the counting rhythm*”. Naturally, no division could be carried out at all.

Approximation of arithmetic results (C22). The patient was asked to point to the approximate result of an operation, without attempting to solve it (e.g. $275 \times 4 = 600, 1200, 2300$ or 50). The results were severely impaired.

Positioning a quantity on a scale (C23-C24). The subject had to evaluate a quantity presented visually ($n = 10$) or auditorily ($n = 10$), on a vertical scale with three tick-marks ranging from 0 (bottom) to 100 (top). For all quantities, the patient placed his correct, although hesitant, responses by comparing them to the reference given in the instructions to the task.

Writing an operation (C25). In this task, the patient was asked to place by column, but not to solve, an operation already written in Arabic code (e.g. $435 + 86 =$). This task revealed no major difficulty.

Written calculations (C26-C28). HP obtained extremely low scores in all problems written in Arabic code, due to systematic errors in the carry-over and to the loss of algorithms in multiplication tasks. For subtractions, his difficulties were mainly due to the fact that he always proceeded by subtracting the smallest from the largest number independently of their respective positions (e.g. lower or upper row).

Perceptive estimation of quantities (C29). Pictures of objects were shown to the patient who was asked to estimate them in terms of quantity, weight, or height. HP was severely impaired for height, weight and number of objects (e.g. the weight of an umbrella was estimated at 10 kg). Occasionally, when HP could compare the object to a known reference, the response was correct. In answering the question: “*how much does this person weigh?*”, HP referred explicitly to his own weight and gave a plausible response.

Cognitive estimation of quantities and precise numerical knowledge (C30-C31). These tasks consist in giving an estimate in a contextually based situation (i.e. “*is 20 pages a lot, a little or normal for a letter?*”) and producing accurate numerical facts (“*how many days in a week?*”). For both tasks, the answers were good.

In order to evaluate the patient’s knowledge about himself and his everyday life, the patient was tested with an encyclopaedic and autobiographic knowledge task composed of twenty-eight different questions (related to his own life: “*What is your license plate number?*”, “*How tall are you?*”; or to everyday life: “*What is the price of a bottle of milk?*”, or “*How many kilometres does a passenger plane cover in one hour?*”). All responses given by HP were considered satisfactory (28/28).

The EC301 suggested that all transcoding tasks involving the Arabic system were impaired, while those involving the verbal/alphabetic system were preserved. This apparent dis-

sociation, however (contradicting McCloskey’s assumptions), could not explain the results in magnitude comparisons (C18-C19). Indeed, the fact that in magnitude comparisons the scores in printed word numbers were inferior to those obtained in Arabic numerals invalidates the hypothesis of a preserved verbal code. These discrepancies may be explained by the fact that reading aloud and writing to dictation can be done only with lexical, phonological and graphemic components, without necessarily referring to the syntactic processes, while this is not the case for magnitude comparisons and transcoding from one code to another.

Considering that the EC301 battery does not specifically evaluate syntactic judgement, we conducted experiments in order to 1) test the hypothesis of an amodal representation of numbers using transcoding tasks according to McCloskey’s model and 2) to evaluate the impact of syntactic components in HP’s scores using a syntactic decision task.

Experiment 1: Transcoding and syntactic tasks

Transcoding tasks

The tasks administered to HP were the following: verbal repetition, reading aloud, pointing (the examiner asked the patient to indicate an Arabic or alphabetic written number among a series of different numbers), writing to dictation both in Arabic and alphabetic codes, transformations from alphabetic to Arabic code (one to 1), and from Arabic to alphabetic code (1 to one). For each of these tasks, the patient was tested with ten items in each of the six following controlled classes: a) units from zero to nine; b) numbers 10 to 19; c) ten numbers representing the tens (e.g. 25); d) ten numbers representing the hundreds (e.g. 534); e) ten numbers representing the thousands (e.g. 1025), and finally f) ten five-digit numbers (e.g. 32246).

Given the fact that the patient’s productions in transcoding tasks were only impaired in the numbers above one hundred, only his scores in the three last categories were retained (hundreds, thousands and ten thousands). Table 4 details the patient’s performances in these categories for each task

Table 4. Transcoding tasks in experiment 1

Tasks	Comprehension system	Production system	Performances
Repetition	Oral verbal	Oral verbal	30/30
Reading	Alphabetic verbal	Oral verbal	28/30
Reading	Arabic	Oral verbal	6/30
			Total: 71%
Dictation	Oral verbal	Alphabetic	25/30
Transcoding	Arabic	Alphabetic	10/30
Pointing	Oral verbal	Alphabetic	30/30
			Total: 72%
Dictation	Oral verbal	Arabic	0/30
Transcoding	Alphabetic verbal	Arabic	1/30
Pointing	Oral verbal	Arabic	19/30
			Total: 22%

as a function of both the input (comprehension) and output (production) modality.

When the scores were considered as a function of the production system, all tasks showed a significant difference between performances in Arabic code (20/90 correct) compared to oral (64/90 correct) or alphabetic (65/90 correct) code. However, it must be noted that, even in the oral and printed number (alphabetic) production systems, his scores were particularly deficient whenever the input involved Arabic code (6/30 correct in reading and 10/30 correct in transcoding). When transcoding from Arabic to alphabetic code, erroneous responses (10/30 correct) comprised 4 errors in the hundreds and 16 errors in the thousands. Of these, 3 errors were lexical (e.g. 64524 transcoded into “*soixante quatre mille deux cent vingt quatre*/sixty four thousand two hundred and twenty four”), and 17 errors were syntactic (e.g. 2612 transcoded into “*vingt six mille douze*/twenty six thousand and twelve”). Among the syntactic errors, there were 6 productions that do not exist in French (890 transcoded into “*quatre vingt neuf cents*/eighty-nine hundred”).

Globally, these results confirmed the advantage of the oral and alphabetic production systems in reading (alphabetical code), dictation and designation. On the other hand, transcoding tasks (Arabic into alphabetic or the inverse), which in addition to lexical, phonological and graphemic components, necessitated also syntactic processes, were impaired and showed a great number of syntactic errors.

Syntactic decision task

In order to determine to what extent such syntactic processes influenced the patient's performances, we designed a syntactic decision task, inspired by the patient's errors. In this task, HP was presented with thirty written alphabetic numbers of which half were comprised of wrong numbers. The fifteen wrong numbers were chosen among HP's own erroneous productions when reading from Arabic code (e.g. “*quatre vingt cent dix huit*/eighty hundred and eighteen”). Each number was presented visually to the patient and was simultaneously read aloud by the examiner. The patient was asked to decide for each item whether it existed in French or not.

The patient scored 18/30 correct responses (60%). He accepted 12/15 numbers among the correct forms and rejected 3 of them (e.g. “*trois mille cent deux*/three-thousand one-hundred and two”). Moreover, he was critical towards only 6 items among the fifteen erroneous stimuli and accepted 9 of them as correct (e.g. “*soixante cinq cent vingt quatre*/sixty-five hundred and twenty-four”). The errors observed suggest a loss of knowledge of the syntactic rules that govern number writing in French in a person who was used to processing number sequences without any difficulty.

The results presented here show that the errors produced by HP in transcoding tasks were syntactic in nature. The fact that the errors were present in both Arabic and verbal alphabetic codes suggests that HP's impairment was not code-dependent, and thus amodal as proposed in McCloskey's model. However, with regard to HP's difficulties in dealing with small

numbers that do not require syntactic processing, we also hypothesised the existence of a semantic impairment. Indeed, the patient was unable to solve very simple problems (Table 3, C20-C21) or to make approximations (C22), although such tasks are usually successfully mastered even by severe aphasics. Our patient's inability to perform approximations may thus result from a specific impairment affecting the analogue magnitude code described by Dehaene (1992) and Dehaene and Cohen (1995). Therefore, in order to verify whether or not the representation of number continuity (according to Dehaene and Cohen) was impaired, we proposed the following experiment.

Experiment 2: Quantity processing tasks

Positioning on a scale

In the EC301 sub-tests C23-C24, HP was able to position a number on a scale only by referring to the example given by the examiner. Here, we asked the patient to place numbers on a straight line with only a starting and an ending point. In the absence of any example and tick-marks on the scale, the patient claimed that he was unable to carry out the task. He stated that he could not carry out the task without knowing what the number represented (e.g. when asked to place the number 25, the patient said: “*twenty-five what? if you tell me what objects you are talking about, I might be able to place them*”). Once a reference was given, the patient then became capable of carrying out the subsequent items.

Verbal numeral fluency

We asked the patient to produce verbally as many numbers as possible below one hundred in one minute. HP produced only 9 items in one minute. Although his scores in the semantic and phonological word fluency tasks were good (see Table 1 and 2), the patient did not even think of simply counting out aloud in order to give more numbers. These numbers (10, 21, 30, 50, 40, 25, 2, 7, 16) were not rank ordered but produced randomly.

Finding the number minus one

HP was asked to subtract 1 from 30 numbers composed of units, teens, tens, hundreds and thousand. All numbers, presented to the patient in Arabic code, were verbalised by the examiner and the patient had to give an oral response. A total of 11/30 correct responses were given. For the units and teens his performance was satisfactory but began to decline in the tens and particularly in the hundreds (e.g. 67 minus 1: the patient subtracted one for each digit of the number and responded 56).

Guessing an unknown number

In this task, the examiner mentally chose a number below one hundred and asked the patient to guess it through closed questions. He was allowed to make a note of the questions and answers in order to avoid forgetting the relevant indications. HP took approximately 90 minutes to reach the correct answer despite the fact that he had shown normal performances

in executive function tests (verbal and figural fluency tasks, Wisconsin Card Sorting test, Luria alternating figures and Stroop test) and other problem solving tasks (such as logical relation tasks, see below). His questions were not targeted enough to approach the solution (e.g. “*Is it bigger than one?*” immediately followed by “*Is it bigger than one thousand?*”). The other questions did not cover the whole range of possibilities (e.g. “*Is it between twenty and thirty?*” and immediately followed by “*Is it between forty and eighty?*”).

The results reported here suggest that the patient was impaired in all tasks referring to the numerical line representation previously described by Dehaene (1992). His inability, to i) mark a numerical position on a scale; ii) evoke numbers in a verbal fluency task (although he performed very well with words), and iii) find within which limits an unknown number was situated, indicated that his semantic representation of the continuity of numbers was severely affected. Although very minor for the units, these difficulties may have greatly influenced his capacity to resolve even very simple problems as previously evidenced (EC 301 protocol). In order to verify to what extent HP’s difficulties in problem solving were related to his capacity to hold a formal reasoning, we conducted this last experiment.

Experiment 3: Formal reasoning tasks

In the first task, referred to as to a “logical relation task” (Rey, 1967), HP was administered 22 semantically and syntactically complex written questions requiring written responses (e.g. “*Qu’est-ce qu’un groupe qui n’est pas formé de ce qui n’est pas animal?*”/name a group not composed of non-animal elements). HP’s scores were normal (percentile 60).

In the second task, inspired from Piaget’s studies (Piaget and Szeminska, 1941), the patient was tested on size relations (relations of transitivity) between three persons. This task was constituted of 15 written questions presented visually (e.g. “*Paul is taller than Henry, but shorter than Louis. Who is the tallest?*”). HP failed on the 15 items. In order to solve this kind of problem, one should either have a mental representation of the constituting elements or graphically (schematically) reconstitute all the elements. HP was unable to perform such a reconstitution either mentally or graphically. In order to

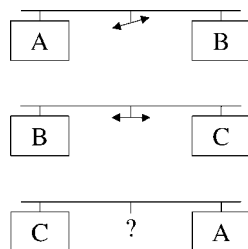


Fig. 2. Schema inspired from Rey (1967) for testing weight relations. Based on the relations at first level (A and B) and the second level (B and C), the patient was asked to predict the relation at the third level (C and A). Although the patient failed to perform a comparable task when the written questions were visually presented, when these questions were presented graphically (schematically), he could show normal performances.

confirm this hypothesis, he was administered a similar task where the items were presented schematically (see Figure 2) in visual form (Rey, 1967). In this case, HP’s performances were normal (percentile 75) suggesting that he could indeed be aided by schematic representations.

Discussion

We report the case of a 59-year-old, right-handed patient (HP), who suffered from acalculia in a context of GS. His impairment consisted of a massive breakdown in numerical abilities in the absence of any symptom of aphasia or intellectual deterioration. The patient was aware of his deficits and could describe the strategies he used to overcome them. Verbal tasks such as reading aloud and writing to dictation in alphabetic code, as well as those based on autobiographic and encyclopaedic knowledge, were well performed. In contrast, numerical tasks, such as reading Arabic numbers aloud, writing to dictation in Arabic code, estimating quantities, comparing magnitudes and performing approximations were severely affected. Moreover, HP was highly impaired in various other tasks including oral counting, dot counting, as well as reciting multiplication tables, which are often assimilated to verbal automatisms. Finally, he was seriously deficient in problem solving, although at the same time, he was able to perform verbal logical relation class tasks.

The hypothesis of a short-term memory impairment was often put forward in the literature to explain the difficulties of non-aphasic patients in dealing with numbers (Dagenbach and McCloskey, 1992; Noël *et al.*, 2001). In the three cases of isolated acalculia due to left parietal lesions reported by Takayama *et al.* (1994), the patients showed no difficulty in transcoding tasks or in spatial alignment. However, they were unable to deal with quantities and to perform the different steps of problem solving. The authors explained such deficits in terms of working memory impairment. Other reports showed that patients suffering from a working memory disorder do not necessarily suffer from acalculia (Butterworth *et al.*, 1996). HP’s poor performance in the digit span (4 digits forward, and 2 digits backward) may be explained by the fact that dealing with numbers, *per se*, constitutes a major difficulty. By contrast, in the various other tasks involving working memory processes, HP’s performances were normal (scores in the “Brown-Peterson” test, repetition of long sentences, etc. see also Mayer *et al.*, 1999). His scores on the Token test, known to correlate with working memory tasks (Butterworth *et al.*, 1996), were excellent. Furthermore, in most of the tasks, the patient had no time limitation and disposed of his own written notes. Consequently, the hypothesis of a working memory impairment does not provide a satisfactory explanation for HP’s deficits.

Since HP’s performances in the EC301 battery (Table 3) and in transcoding tasks were particularly poor in comprehension and production of Arabic numerals, the question of an Arabic code-specific impairment was raised. Nevertheless, subsequent experiments revealed that HP’s errors were not

code-dependent, but occurred also in oral and alphabetic code when the syntactic component was involved. Errors attributable to the syntactic components were present in both transcoding operations (from alphabetic into Arabic code and vice versa, Table 4) and syntactic judgement task. The fact that transcoding tasks for numbers below one hundred were correctly performed, suggests that the access to the internal semantic representation, proposed in McCloskey's model, was possible at least for small numbers. However, we also showed that in the other tasks, where syntactic processing was no longer involved, the patient failed even with very small numeric quantities. Based on this observation, we sought to verify the hypothesis of a deficit affecting the representation of the number continuity. According to Dehaene (1992), this continuity is mentally represented by a numerical line directed from left to right. In order to test the integrity of this representation, four tasks were designed: i) positioning a number on a scale, ii) evoking numbers in one minute, iii) subtracting one from a number and finally iv) guessing a number chosen by the examiner (between 1 and 100). Impaired performances in all these tasks suggested that HP had lost the successive representation of the numbers. These findings fit well with HP's impaired performance in both oral and written counting (EC301, C1-3) but also in other sequential meaningful lexicons.

Given the deficits observed in this case, neither McCloskey's, nor Dehaene and Cohen's models can completely explain HP's semiology. Could HP's difficulties be category-specific? This explanation has previously been proposed for a patient who was impaired in processing numbers above four independently of the modality (Cipolotti *et al.*, 1991). Assuming that numbers that constitute a particular semantic class fall under different headings, as has already been proposed (Hittmair-Delazer *et al.*, 1995; Fuson, 1988), HP's impairments appeared to affect numerical and sequential aspects but not non-numerical ones. For example, HP correctly responded to questions concerning his license plate number or the boiling temperature of water.

The symptoms expressed by HP were only in partial accordance with Dehaene and Cohen's predictions about acalculia in GS (Cohen *et al.*, 2000; Dehaene and Cohen, 1997). In fact, their patient, MAR, who suffered from GS due to a right inferior parietal lesion, was only impaired in quantity manipulation and in problem solving (in the absence of other number processing disorders, Dehaene and Cohen, 1997). This deficit was subsequently designated as a "*semantic anarithmetia*" (Cohen *et al.*, 2000). HP demonstrated similar difficulties, but was also impaired in rote-learning tasks and arithmetic facts, which are thought to rely on verbal automatisms (Cohen *et al.*, 2000; Dehaene, 1992).

Our patient, who presented a particularly pure GS, suffered not only from a "*semantic anarithmetia*" as did patient MAR in Dehaene and Cohen's report (1997), but also from a "*verbal anarithmetia*" as did both BOO (Dehaene and Cohen, 1997) and ATH (Cohen *et al.*, 2000). Accordingly, HP was impaired in the three codes described above

(verbal, visual and analogue (Dehaene and Cohen, 1995). Furthermore, he was totally unable to estimate the result of operations, as opposed to patient NAU (Dehaene, 1992) who was able to do so in spite of a severe aphasia. Taken together, the deficits presented by HP can be referred to as a "*global acalculia*".

The subcortical lesion observed in HP does not implicate the same anatomical network as that predicted by Dehaene and Cohen (1997). They assumed that the direct route, which supports rote arithmetical facts, is composed of a left cortico-subcortical loop through the basal ganglia and the thalamus, as observed in patient BOO who suffered from a capsulo-lenticular stroke. This is not the case for HP, who suffered neither from a thalamic nor from a left fronto-striato-thalamic loop injury. His small subangular lesion is located deep in the inferior part of the angular gyrus and the superior-posterior part of T1 (Mayer *et al.*, 1999). When compared to the lesion in patient MAR, HP's lesion, which isolated the anterior part of the angular gyrus, spared the other regions of the inferior parietal lobule thought to be involved in GS's *anarithmetia*. As previously reported, HP's sub-cortical stroke did not destroy the angular anterior area but rather damaged the connections between this region and the IPL. Based on these anatomical observations and the presumed role of the IPL in GS's *anarithmetia* (Dehaene and Cohen, 1997), one can hypothesize that either the lesion or the isolation of the IPL may be necessary to produce an *anarithmetia* or a "*global acalculia*".

In our previous report, we suggested that the common cognitive denominator of GS was an impairment in the mental manipulation of images (Mayer *et al.*, 1999). A similar explanation has already been advanced by Gold *et al.* (1995) who described a case of GS with an isolated deficit in deriving the relative position of an object along a horizontal axis. However, such an explanation cannot be considered as exclusive for GS given the fact that other neuropsychological dysfunctions, not associated with GS, can also be explained by an impairment in the manipulation of mental images. Actually, Coslett (1998) found that right brain-damaged patients with neglect identified pictures of the contra-lesional left hands significantly less accurately than pictures of right hands. This deficit, suggesting that the identification of pictures of hands involves the matching of the stimuli to a mental representation of one's body, can also be explained by an impairment in the manipulation of mental images.

In our observation, other elements indicate that a deficit affecting the mental representation of images may indeed be at the origin of HP's difficulties. We have already shown that HP's finger agnosia (which necessitates a visual image transformation) was present only when visual support was suppressed and that left-right discrimination was more impaired when the patient had to operate a mental rotation (Mayer *et al.*, 1999). Here, we have in addition shown that while HP experienced major difficulties in problem solving, particularly in "logical relation class tasks" (see experiment 3), he was able to correctly perform such tasks if he was visually aided by a schematic representation of the task elements.

An impairment affecting the manipulation of mental images of spatially related objects could also explain HP's difficulties in number processing and calculation. This impairment was best illustrated by HP's deficits in manipulating number sequences and serial lexicons which are associated with the representation of number continuity, i.e. the so-called "*SNARC effect*" (for "*Spatial-Numerical Association of Response Codes*", Dehaene, 1992). The *SNARC effect* refers to the assumption that numbers are mentally represented in an analogue format and are thus associated with a specific representational space, from left to right in Indo-European languages. An inverted representation, i.e. from right to left, is found in Arabic and other oriental languages (Dehaene, 1992). The spatial nature of this representation has been supported by a recent report showing that the mental number line was disrupted in neglect patients (Zorzi *et al.*, 2002). In this respect, HP experienced great difficulty in simply counting from one to twenty. He resorted to external help such as counting on his fingers and made numerous errors. He experienced difficulties in other sequential or linear lexicons (i.e. he showed a clear increase in response time) such as reciting the days of the week or the months of the year, as well as in positioning a day in a sequence. It should be noted that both numerical lexicons (units, tens, hundreds, thousands) and non-numerical linear lexicons (days of the week, months of the year) are hypothesised to follow a constrained order in the course of their acquisition (Seron, 1993). Impairments in the manipulation of mental images may also affect resolution procedures, particularly when complex operations are concerned. However, since HP failed already in very simple operations, the relation between calculation impairments attributable to the hypothesised deficit in mentally manipulating images, and procedural knowledge could not be formally tested. Concerning the functional independence of number processing/calculation systems and procedural knowledge, one should remember that the emergence of particular dysfunctions such as language impairments (or difficulties in mental image manipulation) may have considerable repercussions on other independent, though intimately linked systems like number/calculation processing.

In conclusion, the patient HP presented here showed a massive, heterogeneous but "*global acalculia*", characterised by a numerical syntactic deficit, difficulties in serial lexicons, loss of arithmetical facts and severe anarithmetia. Considering the fact that HP's deficits were not associated either with a general intellectual deterioration, or with aphasic symptoms, and that the severity of the impairments in number processing and calculation largely exceeded that of the other symptoms constituting GS, we termed the present observation "*pure global acalculia*". In addition, the fact that the deficits affecting number processing exceed the anarithmetia previously described in relation to left parietal lesions and GS, could suggest that the isolation of the IPL might constitute a sufficient condition in producing pure global acalculia.

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Pure global acalculia following a left subangular lesion

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Abstract

We describe the case of a right-handed patient who presented a severe acalculia in the context of a pure Gerstmann syndrome following a subangular lesion that spared the left inferior parietal lobule (IPL). The patient showed impairments in Arabic and verbal codes, in number production and comprehension, as well as in numerical facts and problem solving. By using the EC301 calculation battery, semantic and syntactic tasks in Arabic and verbal codes, we tested the different hypotheses raised by the cognitive neuropsychological models of acalculia. The patients' difficulties, which were not associated with a general intellectual deterioration, and those affecting number processing as a particular semantic class, were indicative of a "global acalculia". This deficit, which exceeded the anarithmetia usually described in Gerstmann syndrome following left IPL lesion, suggested that the isolation of this area may constitute a sufficient condition for producing such a global acalculia. These results are discussed in terms of a disorder in the manipulation of mental images of spatially related objects.

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Acalculia following left subangular lesion

Author's designation of case

Pure global acalculia

Key theoretical issue

- Acalculia in the context of Gerstmann syndrome.

Key words: acalculia; Gerstmann syndrome; left parietal lesion; number processing; calculation

Scan, EEG and related measures

MRI

Standardized assessment

Neuropsychological testing

Other assessment

EC301 number processing and calculation protocol

Lesion location

- Left subangular (sub-cortical parietal) lesion

Lesion type

Ischemic cerebral vascular accident (CVA)

Language

English