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A pure case of Gerstmann syndrome with a subangular lesion

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Summary

The four symptoms composing Gerstmann's syndrome were postulated to result from a common cognitive denominator (Grundstorung) by Gerstmann himself. He suggested that it is a disorder of the body schema restricted to the hand and fingers. The existence of a Grundstorung has since been contested. Here we suggest that a common psychoneurological factor does exist, but should be related to transformations of mental images rather than to the body schema. A patient (H.P.) was studied, who presented the four symptoms of Gerstmann's syndrome in the absence of any other neuropsychological disorders. MRI showed a focal ischaemic lesion, situated subcortically in the inferior part of the left angular gyrus and reaching the superior posterior region of T1. The cortical layers were spared and the lesion was seen to extend to the callosal fibres. On the basis of an extensive cognitive investigation, language, praxis, memory and intelligence disorders were excluded. The four remaining symptoms (finger agnosia, agraphia, right–left disorientation and dyscalculia) were investigated thoroughly with the aim of determining any characteristics that they might share. Detailed analyses of the tetrad showed that the impairment was consistently attributable to disorders of a spatial nature. Furthermore, cognitive tests necessitating mental rotation were equally shown to be impaired, confirming the essentially visuospatial origin of the disturbance. In the light of this report, the common cognitive denominator is hypothesized to be an impairment in mental manipulation of images and not in body schema.

Keywords: Gerstmann's syndrome; MRI; acalculia; mental imagery

Abbreviation: VC = visual control

Introduction

Gerstmann (Gerstmann, 1924) described the case of a 52-year-old woman, who was admitted at the Wiener Psychiatrische Klinik, complaining of memory and writing difficulties. Neurological evaluation showed a right hemianopia, calculation impairment, writing disability, lack of recognition and orientation for the right and left sides of her own body, and loss of ability to recognize, identify, differentiate, name, select and orient the individual fingers of either hand. This latter deficit was called finger agnosia. Gerstmann interpreted the finger agnosia as the selective impairment of the body image in one sphere, 'as though the optic–tactile–kinaesthetic image pertaining to the fingers were split off from the total body image' (Gerstmann, 1924). Three years later, Gerstmann presented two other cases with finger agnosia, left–right confusion, agraphia and acalculia (Gerstmann, 1927). These two patients had no hemianopia but suffered, in addition to the four symptoms, constructional apraxia and impaired colour perception. Gerstmann stated that the association of isolated agraphia with finger agnosia was of great importance, both symptoms being called 'Kardinalsymptome'. The other impairments were described as accompanying deficits ('Randsymptome').

Only in 1930 did Gerstmann, on the basis of other cases, decide to give the symptom complex of finger agnosia, left–right confusion, agraphia and acalculia, the status of a new syndrome, which has since been called the Gerstmann syndrome (Gerstmann, 1930). The phenomenological association of the four symptoms into a single well-defined syndrome was quite significant for the author. For example, the right–left disorientation occurs 'often with special reference to the hands and fingers' (Gerstmann, 1957), the differentiation of fingers is necessary for writing, and fingers play an important role in the first arithmetic operations of children as well as in counting in primitive populations, etc.

Consequently, the basis of the syndrome was related to the existence of a common psychoneurological factor (Grundstorung). Another important assertion was that the syndrome has a high localizing value, pointing to the transition between left angular gyrus to the second occipital convolution.

Emphasis was put on the impairment of the body schema in the interpretation of finger agnosia, even though the finger-localization deficit could also be observed on the examiner's hands. In the same manner, Gerstmann attributed left–right confusion to a change of body image, even though errors were also frequent when the response had to be given on the body of another person. Gerstmann argued that knowledge of one's own body was necessary in order to understand another body.

The idea that an impairment of the body image is the cognitive psychoneurological denominator of Gerstmann's syndrome was very soon challenged. For example, for Herrmann and Potzl (Herrmann and Potzl, 1926), the main factor responsible for the syndrome was 'finger apraxia'. Schilder (Schilder, 1931) even cast doubt on the unity of finger agnosia. He distinguished five types of impairments: finger agnosia (dissolution of the finger schema), finger aphasia (difficulty in naming designated fingers and in indicating fingers named by the examiner), visual finger agnosia (non-differentiation of fingers on one's own hand or another person's, presented visually), apraxia of finger choice (difficulty in imitating finger movements shown by the examiner) and constructive finger apraxia (the incapacity to imitate finger positions given by the examiner, while movements to verbal instructions are preserved). The different forms of finger agnosia were thus characterized by the different ways in which they were elicited (naming versus designation, finger movements, responses on the patient's own hands or on the examiner's, visual open- or closed-loop). He hypothesized a different cerebral localization for each of these symptoms [for a more detailed discussion of this point, see Benton (Benton, 1977)]. In parallel to these attempts at fragmenting the syndrome, other authors tried adding one or more elements to it. Constructional apraxia has been considered by many authors as an integral part of Gerstmann's syndrome (e.g. Stengel, 1944; Hecaen and Ajuriaguerra, 1952). Attempts have been made to relate Gerstmann's syndrome to a broader common denominator. For example, Stengel (Stengel, 1944), describing the case of a woman suffering from a loss of spatial orientation, a constructional apraxia and Gerstmann's syndrome concluded 'constructional apraxia and Gerstmann's syndrome, when occurring in isolation, are incomplete or abortive appearances of the syndrome exhibited by our case'.

A more determined attack against the notion of Gerstmann's syndrome was made by Benton (Benton, 1961). He evaluated seven symptoms with an extensive battery in 100 brain lesioned patients, including 12 patients with left parietal lobe disease. He then established the mean correlation coefficients between individual performances and the syndrome in the 100 brain-damaged patients as well as in the 12 patients with left parietal lobe disease. Results showed clearly that 'the particular combination of behavioural deficits which form the syndrome show no stronger internal associative bounds than do a score of other combinations of behavioural deficits' (Benton, 1961), such correlations as reading versus right–left; – finger localization; – writing; – calculation, being stronger ($r = 0.61$) than, for example, finger localization versus right–left; – writing; – calculation, ($r = 0.38$).

Heimburger and colleagues (Heimburger *et al.*, 1964) presented evidence for one or more components of Gerstmann's syndrome in 111 patients from a population of 465 unselected brain lesioned patients. They concluded that Gerstmann's symptoms had some localizing value in relation to the side of the lesion (78% of the patients presenting the four components of the syndrome had an area of tissue damage strongly lateralized to the left hemisphere versus 9% to the right hemisphere) and to the site of the lesion (among the patients with a complete Gerstmann's syndrome, the left posterior parasylvian area was definitely involved in 57% and not involved in 13%), but they denied the implication of the left angular gyrus in Gerstmann's syndrome, the lesions spreading 'widely over the parietal, temporal, and occipital lobes' (Heimburger *et al.*, 1964). Further, they claimed that 'every patient with four Gerstmann components had associated evidence of severe impairment of brain functions. The syndrome is not to be regarded as an autonomous entity, but merges with numerous other neurological deficits, notably dysphasia' (Heimburger *et al.*, 1964). It should be noted that all of their patients with a complete Gerstmann syndrome were aphasic.

In line with Benton's studies (Benton, 1961) and Heimburger and colleagues (Heimburger *et al.*, 1964), Poeck and Orgass (Poeck and Orgass, 1966) attempted to verify Gerstmann's assertions concerning the isolation of the four symptoms from other behavioural deficits. They examined 50 patients unselected with regard to localization of brain lesion. In addition to the four elements of the Gerstmann syndrome, they tested the presence of dyslexia, impaired verbal memory, constructional apraxia and aphasic disorders. The crucial interest of their study was the demonstration that all of Gerstmann's symptoms, either in isolation or taken together, were strongly associated with aphasia. The authors claimed that 'aphasia might be regarded as the true [Grundstorung] in the Gerstmann symptoms'. They concluded that Gerstmann's syndrome was of little interest for its localizing value, given its redundancy with the localization value of aphasia.

Nevertheless, the existence of Gerstmann's syndrome without language impairment has been shown in numerous studies. Kinsbourne and Warrington (Kinsbourne and Warrington, 1962) described eight patients with the full tetrad of Gerstmann's syndrome, and only two of them showed aphasia. Some case reports (Strub and Geschwind, 1974; Varney, 1984; Sobota *et al.*, 1985; Trillet *et al.*, 1989; Mazzoni *et al.*, 1990; Dehaene and Cohen, 1997; Dozono

et al., 1997) clearly demonstrated the presence of Gerstmann's syndrome without any signs of aphasia. Nevertheless, they all showed other associated disorders. They presented either limb (Dehaene and Cohen, 1997, patient MAR; Dozono *et al.*, 1997), or constructional apraxia (Kinsbourne and Warrington, 1962; Strub and Geschwind, 1974; Sobota *et al.*, 1985; Trillet *et al.*, 1989; Dozono *et al.*, 1997), visual or sensory right-sided deficits (Varney, 1984; Mazzoni *et al.*, 1990), neglect (Dozono *et al.*, 1997) or intellectual deterioration (Kinsbourne and Warrington, 1962; Strub and Geschwind, 1974).

A particularly pure case of Gerstmann's syndrome has been published by Roeltgen and colleagues (Roeltgen *et al.*, 1983). They described a 62-year-old man who presented the four elements of Gerstmann's syndrome without aphasia, alexia, constructional apraxia or memory disorder. This patient was perhaps the unique case who presented a pure Gerstmann's syndrome resulting from a circumbscribed lesion verified by a CT scan. Unfortunately, the evaluation of the four elements was not extensive and did not allow any hypotheses about a potential common cognitive factor underlying the four deficits to be drawn.

In association with other impairments such as aphasia, apraxia or sensorimotor deficits, Gerstmann's syndrome has been described in numerous brain lesions: left parietal, temporal and occipital lobe (Heimburger *et al.*, 1964), left frontal posterior (Brusa *et al.*, 1960), left thalamic (Santos *et al.*, 1991) and right parietal in left-handed patients (Moore *et al.*, 1991; Dehaene and Cohen, 1997; Dozono *et al.*, 1997). However, when accompanying deficits were moderate or absent, Gerstmann's syndrome was produced by left parietal disease in right-handed patients (Roeltgen *et al.*, 1983; Varney, 1984; Trillet *et al.*, 1989; Mazzoni *et al.*, 1990). Moreover, the CT scan of Roeltgen's patient showed a focal 'cortical lesion of the superior angular gyrus, that extended into the supramarginal gyrus and minimally into the superior parietal lobule'. Morris *et al.* (Morris *et al.*, 1984), in their cortical stimulation study, produced an isolated Gerstmann's syndrome by stimulating an area located in the transition between the angular and supramarginal gyri. They made the hypothesis that if a pure Gerstmann's syndrome were to be found, the lesion would most likely be small and limited to the region of the angular gyrus.

In order to analyse the existence of a common psychoneurological factor in Gerstmann's syndrome, a number of prerequisites must be obtained. First, the four elements of the syndrome must be present. Secondly, the evaluation must exclude any neurological or neuropsychological deficits which could interfere with the syndrome, e.g. the patient must not suffer from motor or sensitivity impairments, visual defect, language, praxia, gnosia, memory or intellectual impairment. Thirdly, given the multifactorial aspects of the four symptoms, each one must be studied in detail, e.g. observing input and output channels and verbal/non-verbal aspects of the task, etc.

Here we report the case of a patient who, following a circumbscribed cerebral vascular accident, presented the four elements of Gerstmann's syndrome without any signs of aphasia, apraxia, amnesia or intellectual deficit. An experimental approach was adopted to study the characteristics of the tetrad.

Method and results

Case report

H.P. is a 59 year-old right-handed man with university education (degree in literature), who was employed as an insurance agent until his illness. On February 28, 1994, he suddenly became unable to write, calculate or dial telephone numbers. He came to the emergency ward the same day. The patient was alert and oriented. The cognitive defects are described below. The remaining neurological evaluation was within normal range: notably there were neither visual nor motor deficits. Sensation for pain and temperature were normal, as were vibration sense and kinaesthesia. Cerebral CT scan revealed a small left posterior parietal ischaemic lesion. He remained in the neurological ward for 10 days. No embolic source could be found and the patient was discharged with antiplatelet medication . He did not show any further sign of stroke or other neurological symptomatology during the follow-up. Clinical data reported here were collected in the 3 months post-onset. No evolution of the symptomatology was found during this period. H.P. gave written consent to participate as did all control subjects. The study was approved by the ethical committee of the Geneva University Hospital.

Neuroradiological study

Two successive MRI scans at 3-year intervals confirmed a focal posterior subangular lesion involving the posterior part of the angular gyrus, consistent with ischaemia revealed at the CT scan. The first MRI showed a small posterior inferior parietal subcortical lesion, immediately lateral to the temporo-occipital ventricular horn (Fig. 1A). There was a subcortical extension towards the superior parietal gyrus (Fig. 1B).

The basic neuroanatomical study is based on the second MRI, 3 years after the stroke. This latency allowed a better visualization of the secondary degeneration or atrophy. A special template reconstruction was drawn directly during the MRI procedure, based on a stereotaxic method (Talairach and Tournoux, 1988). These MRI slices were obtained with the brain in the bicommissural plane with horizontal T_2-weighted slices and sagittal T_1-weighted slices. Additional 3D T_1 sequences were obtained and reconstructed in the bicommissural horizontal and coronal planes. Lesion site was established independently by a neuroradiologist and a neurologist using the horizontal and coronal plates of the Talairach–Tournoux atlas adapted to MRI procedures. Both examiners arrived at the same conclusion. The site is subcortical, corresponding to the horizontal plate, 50 mm

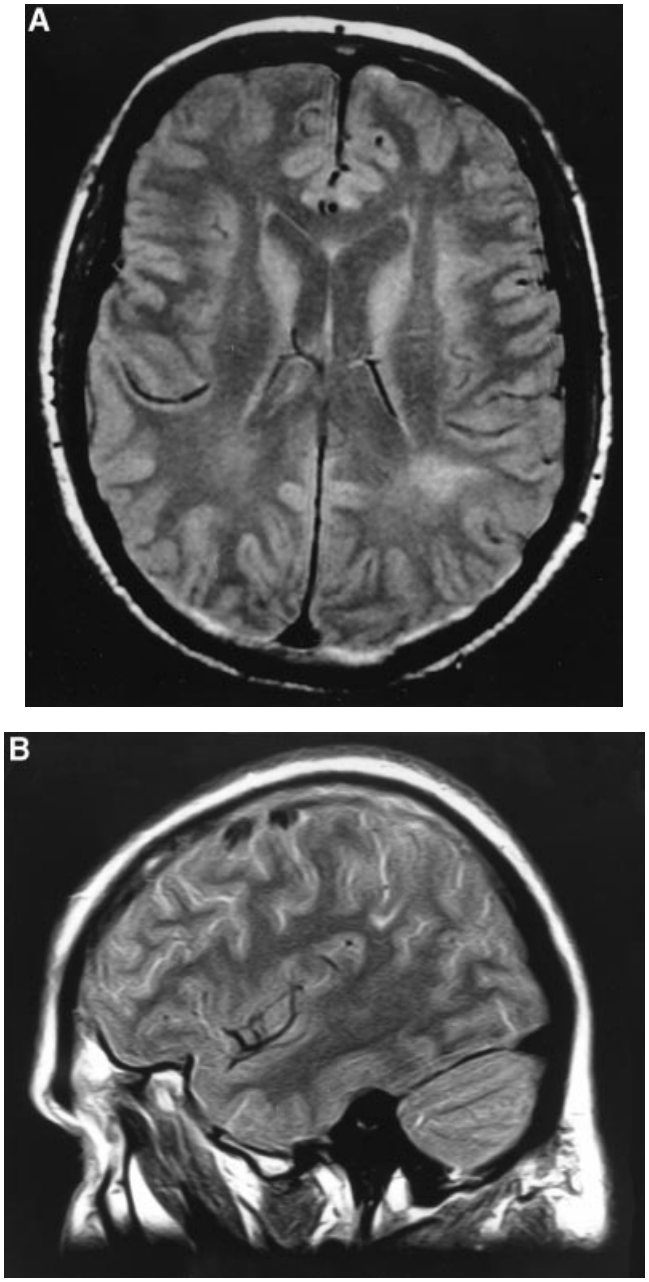

Fig. 1 MRI axial (**A**) and sagittal (**B**) sections of H.P.'s brain showing the left subangular ischaemia.

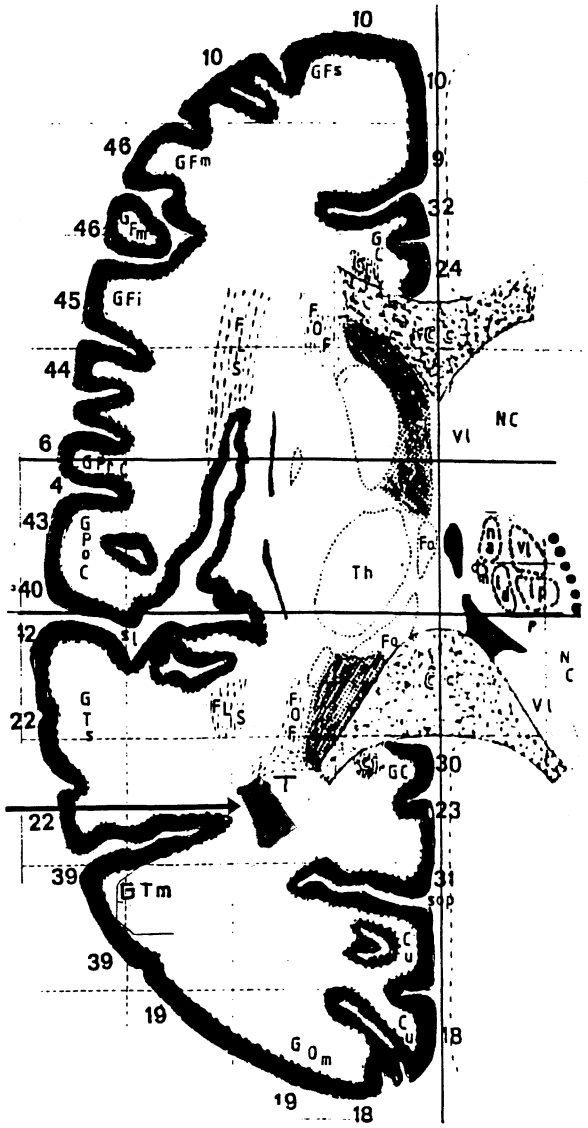

Fig. 2 Axial schematic plate according to the atlas of Talairach and Tournoux (Talairach and Tournoux, 1988) showing the subcortical lesion (arrow) and its topographic relationships with the angular gyrus (area 39).

posterior to the anterior commissure. This corresponds to a small subcortical lesion situated under the lower and anterior part of the angular gyrus, the upper and posterior part of the superior temporal gyrus, interrupting associative fibres from the angular gyrus to the superior parietal area and to the contralateral homotopic areas. Anteriorly, it reaches the caudal limit of the supramarginal gyrus (Fig. 2). Moreover, we were able to observe a small subcortical extension of the lesion towards the superior parietal lobule, with some corresponding cortical atrophy at this level. Finally, a secondary atrophy of the splenium of the corpus callosum could be observed. Thus, the lesion met the criteria given by Greenblatt (Greenblatt, 1976), in order to be qualified as subangular, i.e. subcortical and ventral to the angular gyrus. Nevertheless, its localization was slightly posterior and dorsal to those of the original description.

Neuropsychological examination

H.P. was well oriented and co-operative. His insight into his difficulties was particularly good, and helped us to guide our investigation. He scored at centile 85 on the Raven progressive matrices. He obtained a full-scale IQ of 89, a verbal IQ of 97, and a performance IQ of 79 on the Wechsler Adult Intelligence Scale—revised. Individual age-scaled scores are given in Table 1.

The patient scored within the normal range on the Wechsler Memory Scale—revised obtaining the following quotients:

Table 1 *Wechsler Adult Intelligence Scale showing individual quotients and subtest scores*

Subtest	Subtest score
Verbal IQ	97
Performance IQ	79
Information	14
Digit span	3
Vocabulary	13
Arithmetic	3
Comprehension	14
Similarities	12
Picture completion	8
Picture arrangement	7
Block design	5
Object assembly	8
Digit symbol	4

verbal: 119; visual: 106; general memory: 114; attention and concentration: 51; and delayed memory: 123. Long-term memory was also tested with the Rey Auditory–Verbal Learning Test and the Rey Visual Design Learning Test. The patient scored at centile 60 in both tests. Reproduction from memory of the Rey's complex figure was good (centile 80). Buccofacial praxis was unimpaired. Limb praxis was assessed by asking the patient to perform a variety of actions both on verbal command and imitation (described in Hecaen, 1978) and was found to be normal. No constructional apraxia was elicited either when copying 3D drawings and the Rey–Osterrieth complex figure, or when assembling sticks in order to reproduce a cube. No visual agnosia was present [maximal score on the Montreal–Toulouse battery for visual agnosia (Agniel *et al.*, 1992)], but H.P. was deficient (13/30 correct responses) in the Hooper Visual Organization Test (Hooper, 1985). Neglect was assessed using a protocol habitually used in our clinic which comprises cancellation tasks and line bisection, graphic motor productions and descriptions of well-known places. On a test similar to Albert (Albert, 1973), H.P. performed at maximum level. No deviation from the centre was elicited in line bisection. Description of the 'Place Neuve' (a public square in the heart of Geneva) was homogeneous from two opposing imagined points of view, containing details from the left and right side of the imagined space. The indication of body part (except for fingers) on verbal command and on tactile stimulation was perfect (16/16 in both tests). Tactile gnosis was perfect (16/16 objects recognized with both hands). He scored within the normal range on the Wisconsin Card Sorting Test.

Spontaneous speech was fluent. H.P. underwent the French version of the Boston Diagnostic Aphasia Examination (BDAE) (Goodglass and Kaplan, 1972; Mazaux and Orgogozo, 1981). Oral naming, apart from numbers, was perfect [90/90 in a French naming test (Bachy-Langedock, 1989)]. Repetition of high and low probability sentences and oral comprehension subtests were flawless. Verbal fluency was good: 20 animals in 1 min. H.P. scored 10/10 on oral reading and paragraph comprehension. In an abridged version of the Token Test (De Renzi and Faglioni, 1978), H.P. scored 35/36. Performances on specific writing tasks are described below.

In summary, a selective impairment was shown in both subtests of the Wechsler Adult Intelligence Scale—revised implying calculation and number processing (digit span and arithmetic). The verbal IQ prorated without digit span and arithmetic was good (114). In the performance IQ, two subtests (digit symbol and block design) showed very low scores (4 and 5). A closer analysis showed that in both tasks, the patient was very hesitant and in one instance, a right–left inversion was observed. Slow performances in the block design subtest and poor results on the Hooper test pointed to difficulties in mental imagery and rotation, but the possibility of a global intellectual impairment was excluded. The deficient scores in the Wechsler Memory Scale—revised subtests concerning short-term memory, attention and concentration were due to calculation and number processing difficulties. Moreover, there were no other signs of aphasia, apraxia, agnosia or amnesia.

The four elements of the syndrome

Finger agnosia

As Schilder (Schilder, 1931) stated, finger agnosia is not a unitary disability (for a recent review, see Benton and Sivan, 1993), but regroups a certain number of different performances depending on the way it is tested. The nature of the stimulation is important, as well as the mode of response. We tested H.P. in nine situations which combined three types of stimulation (verbal, tactile, visual) and three modes of response (verbal, pointing on one's own hand, or pointing on diagram of a hand). A condition in which vision of a diagram of a hand was allowed (VC for visual control) was compared with the same condition in which no visual feedback of the hand was permitted (without VC for no visual control). Moreover, the hypothesis of Conrad (Conrad, 1932), who saw finger agnosia as an inability to dissociate the parts of a whole ('Ganzheitstorung'), was tested with Kinsbourne and Warrington's (Kinsbourne and Warrington, 1962) procedures. In order to test Gerstmann's hypothesis relative to a circumscribed dissolution of the body schema affecting the fingers, we added an evaluation of finger agnosia on the foot ('toe agnosia').

Testing the input and output modalities. Situation 1 (verbal–verbal): the subject was asked to indicate which finger was used in different situations. For example, he was asked which finger is used to indicate direction, to hitch-hike, on which finger a wedding ring is placed, which finger is the biggest, the smallest, etc. Out of a total of 10 questions, no errors were made.

Situation 2 (verbal–pointing to one's own hand without VC): the subject was asked verbally to point to the index, middle, etc. of either hand out of vision. Five stimulations

Table 2 *Digital agnosia: number of errors in each situation (S1 to S9)*

Responses	Verbal	Pointing to own hand without VC	Pointing on a diagram of a hand with VC
Stimulation			
Verbal	S1: 0/10	S2: 8/50	S3: 0/50
Visual	S4: 0/50	S5: 4/50	S6: 0/50
Tactile	S7: 15/50	S8: 8/50	S9: 4/50
Total	15/110	20/150	4/150

were produced on each finger in a pseudorandom order. Five errors were produced on the right hand and three on the left.

Situation 3 (verbal–pointing on a diagram): the subject was asked verbally to point to the index, middle, etc. on a diagram of a left and right hand. Five stimulations were produced on each finger in a pseudorandom order. No errors were made.

Situation 4 (visual–verbal): the patient laid his hands on the table in front of him. Each finger of both hands was designated without being touched five times in a pseudorandom order. The patient had to state verbally which finger was designated. No errors were elicited.

Situation 5 (visual–pointing on one's own hand without VC): both of the patient's hands were laid on top of a diagram of a hand. The experimenter pointed to a finger on the sketch and the patient had to point to the same finger on his own hand without seeing it. Each finger of both hands was designated five times in a pseudorandom order. Two errors on the right hand and two on the left were elicited.

Situation 6 (visual–pointing on a diagram): both hands of the patient laid on top of a diagram of a hand. The experimenter pointed to one of the patient's fingers without touching it and he was asked to point to the same finger on the diagram. Spontaneously, the patient used a strategy which consisted of applying directly the pointed finger to the corresponding one on the diagram. No errors were seen.

Situation 7 (tactile–verbal): out of vision, each finger of both hands was stimulated five times in a random order. The patient had to give the name of the finger that was touched. Six errors were made on the right hand and nine on the left hand.

Situation 8 (tactile–pointing on one's own hand without VC): behind a screen, each finger of both hands was stimulated five times in a random order. The patient was asked to point to the stimulated finger. Four errors were made on the right hand and four on the left hand.

Situation 9 (tactile–pointing on a hand's sketch without VC): each finger of both hands was stimulated five times in a random order. The patient had to point to the same finger on the diagram of the hand. Two errors on the right hand and two on the left were elicited.

In all these situations no errors were elicited on the thumb or the little finger. The total number of errors for both hands in all situations is presented in Table 2. Comparative analysis of responses showed that vision played a crucial role in performance. Prevention of vision in input or output modality (S2, S7, S8) led to an increase in the total number of errors (31/150) compared with the other situations (S3, S4, S5, S6, S9) where vision was allowed either for input or for output (8/250) [$\chi^2(1) = 32.5$, $P > 0.001$].

Testing the integration. In the next situation the procedures are the same as those of Kinsbourne and Warrington (Kinsbourne and Warrington, 1962).

Situation 10: two fingers are simultaneously touched and the patient was asked to indicate the number of fingers between the ones touched (inbetween test). The 10 pairs of stimulations were carried out three times. Out of 30 responses, H.P. made 10 errors on the right hand and 10 errors on the left.

Situation 11: the fingers were touched in two places. The patient had to determine whether the two stimulations were on the same or on different fingers (two-point finger test). For each hand, 12 double stimulations were carried out on the same finger and 12 on separate fingers. Out of 24 responses, seven errors were made on each hand. All the errors were 'same' responses although two different fingers were touched. The errors were distributed between the index, the middle and the ring finger; no errors were elicited when the little finger was stimulated.

Situation 12: the fingers were touched in two places, either by one matchbox slipped between two fingers or by two matchboxes each touching a different finger. The patient was asked to decide whether he was being touched by one or two matchboxes (matchbox test). The nine different stimulations possible have been applied three times. H.P. did not produce any errors.

Situation 13: four wooden blocks were designed around which the subject was asked to wrap his fingers. The forms of the blocks were such that the patient's fingers were forced into a given pattern. He was then asked to select the corresponding block from among four models (finger block test). The four blocks were used three times in a random order. Out of 12 trials with each hand, H.P. did not make any errors.

Situation 14: five strips of paper on which the names of each finger were inscribed were read by the patient. He was then required to arrange them in order corresponding to the fingers on a hand (finger strip test). The strips were presented five times. On the first trial, H.P. placed the thumb, the ring, the middle, the little and the index fingers, immediately saying: 'no, it's wrong', and correcting the positions. He then placed the strips correctly on the next trials.

Situation 15: the patient and the examiner were seated face to face with their hands on the table, palms facing down. The patient could not see his own hands. The examiner would raise a finger and the patient was asked to imitate the examiner. All fingers of both hands were raised five times in a random order. H.P. made nine errors with his right hand and eight errors with his left. No errors were observed with the thumb and little finger.

Testing toe-agnosia. Situation 16: patient's toes were stimulated without VC. The patient was asked to give the number corresponding to each toe (1 for the big toe, 2 for the second and so on). Out of 25 trials with each foot, 10 errors were produced with the right foot and 11 with the left. No errors were made on the two extreme toes (1 and 5). Situation 17: patient's toes were stimulated without VC. H.P. was asked to point to the stimulated toe on a diagram of a foot. Out of 25 trials with each foot, six errors were produced with the right foot and nine with the left. Only one error was produced on an extreme toe (the fifth toe of the left foot was mistaken for the fourth).

Five neurologically normal subjects, matched for level of education (university degree), sex and age (53, 62, 62, 63 and 70 years), carried out the finger and toe agnosia tests for comparison purposes. A ceiling effect was observed on all tasks. For finger agnosia, no errors were made in any condition by any of the subjects. For toe agnosia (situation 16), the 25 trials yielded an average of 1.6 errors with the left foot (range 0–3) and 1.4 with the right (range 0–3). Situation 17 showed a similar ceiling effect with 1.4 (range 0–3) and 1.2 (range 0–2) for the left and right foot, respectively. The performance by controls being well above that of H.P., statistical comparison was deemed unnecessary.

Summary. The perfect performance in situations 1, 3 and 4 show that finger agnosia was not due to verbal comprehension or production impairment, when vision was allowed. Similarly, no errors were found in tasks implying verbo–visual (S3) or visuo–verbal (S4) transformations. H.P. made no errors in situations where he could see his fingers (S4) or a diagram of a hand (S3 and S6). The difficulties cannot be attributed to an impairment in touch sensitivity (he performed perfectly at the matchbox test), or to an impairment in proprioception (no errors were produced at the finger block test). Moreover, finger agnosia was also present for the toes. The impairment was particularly important when the situation required an internal visual representation of the hand without visual support, independently of whether the entry was verbal (S2) or tactile (S7 and S8) and in the inbetween or two-point finger test.

Right–left discrimination

As for finger agnosia, right–left discrimination is a very broad concept depending on verbal, sensory, conceptual and visuospatial factors (Benton, 1977; Denes, 1989). Following Benton and Sivan (Benton and Sivan, 1993), this judgement requires the analysis of operationally defined components such as orientation with respect to one's own body with or without visual guidance, orientation with respect to an examiner or picture facing the patient, or combined orientation with respect to one's own body and a person in front of the patient.

On the Benton form (Benton, 1959) H.P. scored 22, which is clearly below the norm (mean = 31.2; SD = 1.6). At the Culver form (Culver, 1969) the number of correct responses was 15, which is within normal limits (mean = 17.16; SD = 2.89). Nevertheless, on each trial, H.P. hesitated, looked at his hands or feet, turned them, etc., before giving his response. This markedly slowed his response time (355 s for the entire test, the mean and the standard deviation in a normal reference population being 40.8 and 16.2, respectively). In order to assess the influence of factors such as visual versus non-visual guidance, simple versus complex orders, and congruous versus non-congruous positions of a model, a number of control conditions were added. (i) Under VC, an oral command was given asking the patient to point to his right or left ear, eye, cheek, shoulder, eyebrow, thigh, knee and foot (the same organ was never designated twice running). The order was determined in a pseudorandom fashion and no instruction was given concerning the hand with which the patient should point. The number of correct responses was 15/16. (ii) The procedure was the same as situation 1, but with a two-stage command (the hand with which to point was specified). The number of correct responses was 14/16. (iii) The procedure was the same as situation 1, but the patient was blindfolded. The number of correct responses was 14/16. (iv) Same procedure as in situation 2, but the patient was blindfolded. The number of correct responses was 12/16. (v) Same procedure as in situation 1, but pointing had to be made on a line-drawing model of the body facing the patient (incongruous position). The number of correct responses was 14/16. (vi) Same procedure as in situation 2, but pointing had to be made on a line-drawing model of the body facing the patient (incongruous position). The number of correct responses was 11/16. (vii) Same procedure as in situation 1, but pointing had to be made on a line-drawing model of the body facing away from the patient (congruous position). Eyes and eyebrows, not visible in this position, were replaced by hips and calves. The number of correct responses was 16/16. (viii) Same procedure as in situation 2, but pointing had to be made on a line-drawing model of the body facing away from the patient (congruous position). Eyes and eyebrows, not visible in the pronation position, were replaced by hips and calves. The number of correct responses was 14/16.

The results are summarized in Tables 3A and B. They show a tendency towards an increase in errors depending on the complexity of the instructions, congruity of the examiner's position and presence or absence of visual control. We retrospectively classified the different items with respect to the number of mental processes necessary to resolve the task. For example, deciding which hand has to respond corresponds to one process (1P), deciding which part (left or right) of the body has to be pointed to requires another process (2P) and mentally rotating the body image (in case of incongruous position of the examiner) a third process (3P). In 1P tasks, H.P. made three errors out of 58 occurrences, in 2P tasks 12 errors out of 64 occurrences, and in 3P tasks, five errors out of 16 occurrences [$\chi^2(1) = 7.7$, $P = 0.02$].

The five control subjects also carried out the right–left

Table 3
(A) *Right–left discrimination: number of errors following simple and complex orders with and without visual control (*n = *16 in each situation)*

	One-stage commands	Two-stage commands
With vision	1	2
Without vision	2	4

(B) *Right–left discrimination: number of errors following simple and complex orders with congruous versus incongruous positions of the model (*n = *16 in each situation)*

	One-stage commands	Two-stage commands
Congruous position	0	2
Incongruous position	2	5

disorientation tests. No errors were seen for any subject in any situation.

Summary. The results in Benton's (Benton, 1959) and Culver's (Culver, 1969) tests showed a global impairment in right–left discrimination. Additional tests suggest a role of factors such as complexity of the instruction, possibility of visual guidance and congruity of the model (facing towards or away from the patient). The number of mental processes necessary to solve the different tasks seem to influence H.P.'s performance.

Dysgraphia

Three types of agraphia are classically described: (i) aphasic agraphia, whose symptoms are paragraphias, simple omissions and substitutions or jargonagraphia but with well-formed letters, due to left temporal or subcortical hemisphere lesion; (ii) apraxic agraphia, characterized by a defective form of the letters and in the most severe case a scrawl, seen in most cases after left parietal lobe lesions, and (iii) spatial agraphia consisting in an erroneous management of space on the paper, with respect to either the horizontality or the margin, observed more often following right parietal lesion (Roeltgen and Heilman, 1985). Cognitive neuropsychological models allow a distinction between linguistic processes committed in writing from the realization itself. They separate central processing with phonological and lexical components (Caramazza, 1988) from peripheral modality-specific processing (Ellis, 1982).

In relatively pure Gerstmann's syndrome, it is interesting to note that a peripheral dysgraphia is often present (Martory, 1996). On the contrary, central agraphia (with a phonological or lexical disturbance) is never observed (Lang, 1994) .

H.P.'s writing impairment has been described elsewhere (Zesiger *et al.*, 1997). The patient complained about his handwriting which he reported as being slow and resembling that of a child. He could easily produce sentences, without spatial dysgraphia. Spontaneous written production and writing to dictation were characterized by systematic confusions between lower case letters b and p and between d and q. [*dame*] (lady) was written [qame], [*qui*] (who) was written [dui] and so on. The substitutions concerned exclusively a top-bottom letter reversal and not a left–right shift. H.P. could correctly write all the letters of the alphabet in upper case. He could also produce the lower case letters, with the exception of b–p–d–q. Some letter omissions appeared in the middle or at the end of long words.

Apart from the BDAE, written spelling was assessed with the *Batterie cognitive d'examen de l'ecriture* (De Partz, 1994). H.P. was asked to write 312 words from dictation. Error analysis showed that there was no effect of syntactical class, orthographic regularity, degree of imagery and frequency of the stimuli. Of the 312 words of De Partz's battery, 158 contained p–b–q–d of which 149 were incorrectly produced. Mental imagery was good for all letters apart from p–b–q–d, whose description corresponded exactly to the written substitutions. A deficit of allographic level was hypothesized. Conversely, H.P.'s proficiency in shorthand seemed unaffected by the damage and he used it spontaneously when writing. He was asked to produce the 26 letters of the alphabet both in lower and in upper case, 10 digits, and 20 words in shorthand both with the right hand and with the left hand. On the basis of the results, it was concluded that motor representations for shorthand symbols were preserved and conversely, motor representations for some letters and digits were impaired (Zesiger *et al.*, 1997).

Summary. H.P. presented a peripheral dysgraphia with a moderate disturbance of the allographic level and a strong deficit of the motor graphic patterns.

Dyscalculia

Hecaen *et al*. (Hecaen *et al*., 1961) distinguished three types of mathematical disabilities: (i) aphasic acalculia whose origin lies in a language disturbance and corresponds to a left hemispheric lesion; (ii) spatial acalculia which is an incapacity to organize numbers in space in order to carry out operations and whose lesion is in the right hemisphere; (iii) anarithmetia corresponding to a loss of arithmetical procedures and occurring after bilateral lesions or accompanying dementia.

MacCloskey and Caramazza (1987) constructed a cognitive model of number processing based on linguistic analogies. They distinguished comprehension, production, and calculation systems as well as a long-term memory component. Comprehension and production systems were divided into arabic and alphabetic components. Arabic components, which appeared only in writing modality, were processed through the syntactic and semantic levels. The alphabetic component was further processed at a phonological

Table 4 *Acalculia: number of correct responses in transcoding tasks*

	n	*n* correct
Repetition of numbers (phonological aspect)	12	12
Reading alphabetic numbers	12	12
Reading arabic numbers	12	6
Writing alphabetic numbers under dictation	12	12
Writing arabic numbers under dictation	12	4
Writing arabic numbers given alphabetic numbers	12	6
Writing alphabetic numbers given arabic numbers	12	2
Perceptual estimation of quantities	12	4
Cognitive estimation of quantities	10	10

level. Finally, an abstract modality, the semantic representation, bound these three systems together.

The patient presented difficulties when counting or carrying out operations. When counting from 1 to 20, H.P. had to count on his fingers. This would be done very slowly, sometimes over 5 s from one number to the next (for example when passing from 9 to 10). Furthermore, in the same series of 1–20, three numbers were omitted. When asked to count 3 in threes (starting at 1), the patient was unable to begin the task, even when allowed to use his fingers. The difficulty appeared to be due to the process of incrementing a value along a continuum.

Every task requiring numbers was severely impaired. He was examined using the Calculation and Number Processing Battery developed by Deloche and Seron (Deloche and Seron, 1991). This test contains 13 tasks examining all aspects of number knowledge and calculation and particularly transcoding tasks with reference to MacCloskey and Caramazza's model. Results from the transcoding tasks are shown in Table 4. Tasks using the alphabetic code were all preserved, confirming the absence of language disturbances. Conversely, tasks using the arabic code were impaired.

The patient could write down an operation with no difficulty but was unable to solve it. Moreover, he could not make any estimation about the results of the operation. He also failed in quantity comparisons using arabic and alphabetic codes. Moreover, when asked about the size or the weight of drawings of objects, his perceptual estimation of quantities was wrong. For example he could not estimate the size of a plant even when it was drawn next to a chair. Some abilities were normal such as the cognitive estimation of quantities and precise numerical knowledge (e.g. the usual number of passengers on a bus). This cognitive ability was significantly better than perceptual estimation [$\chi^2(1) = 10.48$, $P = 0.001$].

Summary. H.P.'s acalculia did not correspond to an aphasic acalculia since all tasks requiring language processing, namely reading and writing, were preserved. In accordance with MacCloskey and Caramazza's model (MacCloskey and Caramazza, 1987), we hypothesize an impairment of the arabic system both for comprehension and production. Furthermore, H.P. was significantly impaired in estimating quantities from visual information.

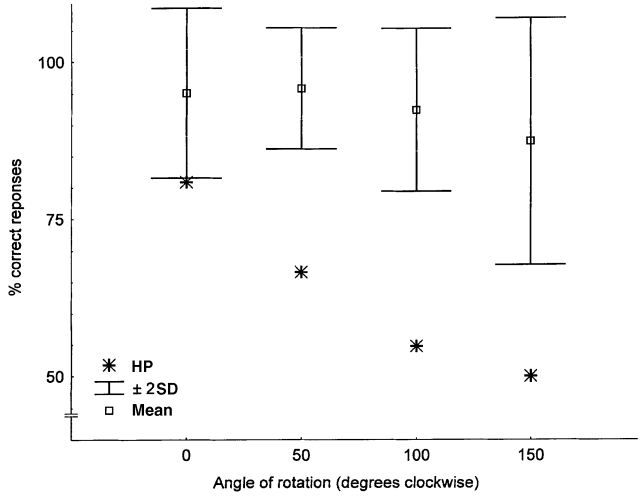

Fig. 3 H.P.'s and controls' performances in the mental rotation task (mean and 2 standard deviations for control subjects are given by open squares and vertical bars, respectively).

Mental rotation

Since the bulk of data found in the different tests suggested difficulties in manipulation of mental imagery, we tested explicitly this hypothesis by giving the patient a mental rotation task.

We applied a computerized version of the Shepard and Metzler (Shepard and Metzler, 1971) mental rotation paradigm, using alphanumeric stimuli (Pegna *et al.*, 1997). In this task, a pair of alphanumeric stimuli were presented on a computer screen. The reference, a normal upright stimulus, was followed after 1050 ms by the test stimulus (a mirror or normal version of the reference rotated by 0, 50, 100 and 150°, clockwise). The patient had to determine if the test stimulus was mirror or normal compared with the reference stimulus (see Pegna *et al.*, 1997, for more detail). An abridged version of the test, used for clinical purpose, was applied to H.P. Twenty-one trials were applied for each angle tested. The number of correct responses was recorded and compared to those of a control group described in the previous study. Figure 3 shows that, compared to that of controls, H.P.'s performance is at the lower limit (as defined by 2 SDs from mean) when no mental rotation is required and that his performance decreases proportionally with the increase in angle of rotation. These results point to an impairment in manipulation of mental images.

Discussion

Patient H.P. provided a unique opportunity to study Gerstmann's syndrome. First, he presented all four elements of the syndrome. This configuration is not very common, as

demonstrated by group studies (Heimburger *et al.*, 1964). Secondly, he did not show other neurological or neuropsychological deficits. In particular, he did not suffer from any visual, sensory or motor deficits, or show any language, praxia, gnosia, memory or general intellectual impairments. Until now the only case showing Gerstmann's syndrome without any other deficit was that of Roeltgen and colleagues (Roeltgen *et al.*, 1983). Thirdly, the cerebral lesion was very circumscribed, confirming Morris and colleagues' (Morris *et al.*, 1984) hypothesis that if a pure Gerstmann's syndrome were to be found, the lesion would most likely be small and situated in the region of the left angular gyrus. Therefore, our results are in accordance with one of Gerstmann's hypotheses concerning the localizing value of the syndrome (see also Benton, 1992).

The existence of a common psychoneurological factor which could explain the Gerstmann symptomatology has been suggested since the first description of the syndrome. However, none of the hypotheses put forward was entirely satisfactory concerning H.P. For example, Heimburger and colleagues (Heimburger *et al.*, 1964) attributed the origin of Gerstmann's syndrome to widespread intellectual impairment. This hypothesis can be excluded in our case, H.P. scoring well within the norms in intellectual assessment measured by Progressive Matrices and Wechsler Adult Intelligence Scale—Revised. The idea expressed by Poeck and Orgass (Poeck and Orgass, 1966) that aphasia could be the 'Grundstorung' of Gerstmann's syndrome can be rejected, all language tests being correctly performed with the exception of writing productions. Stengel's explanation attributing Gerstmann's syndrome to a larger syndrome including loss of spatial orientation and a constructional apraxia (Stengel, 1944) cannot be retained since such deficits were absent in our case. Gerstmann (Gerstmann, 1957) claimed that the common psychoneurological factor of the tetrad was a selective disorder of the body schema in one area of the body, the hand. He wrote: 'As far as all the other parts of the body (including the feet and the toes) are concerned, ability to recognize them and to orient with reference to them remain essentially unaltered'. We tested explicitly this assertion with H.P., and found a toe-agnosia (i.e. finger agnosia for the feet). Interestingly, this type of impairment was limited to the medial fingers as specifically observed for the hands in our patient and others (Gerstmann, 1927; Kinsbourne and Warrington, 1962). This observation leads to the hypothesis that the deficit is common for the hands and the feet. Moreover, the presence of toe agnosia excludes Gerstmann's hypothesis of a selective disorder of the body schema in one sphere. It also excludes the assertion put forward by Critchley (Critchley, 1966) who suggested that the localized autotopagnosia could be due to the unique role of the hands in human activities.

Impairment of specific visual integrative processing has been postulated to take into account all four cardinal components of Gerstmann's syndrome. Levine *et al.* (Levine *et al.*, 1988) and Lang (Lang, 1994) put forward the hypothesis that in Gerstmann's syndrome, the basic psychoneurological problem was defective visuospatial–language integration. Therefore, 'showing a finger in response to its dictated name requires performing a spatial analysis in response to a linguistic stimulus, similar to writing a letter of the alphabet to dictation'. Analysis of H.P.'s errors when testing for finger agnosia did not confirm this assertion: he performed perfectly in tasks where, on verbal instruction, he had to point to a finger, or one on a diagram, under visual control. Finger agnosia was found both in sensory and verbal modalities. Number production was similar in both digital and verbal modalities. Consequently, functional verbovisual dysconnection cannot be implicated in H.P.'s performances.

Another possible visual integrative defect was suggested by Gold and colleagues (Gold *et al.*, 1995) who described a case with an isolated deficit in deriving the relative position of an object along the horizontal axis. They postulated that defective horizontal mapping could account for the other components of Gerstmann's syndrome. We did not specifically test this hypothesis with our patient, but the fact that his writing errors concerned exclusively a top-bottom letter reversal and not a left–right shift does not confirm this idea as a general primary defect.

A fundamental link between the four symptoms could be the difficulty of H.P. in transforming visuospatial images. Such a hypothesis is compatible with the fact that H.P. performed poorly in the Hooper and Kohs' tests. Clearly, more basic visuoperceptual disorders, such as neglect or simultagnosia, may also provoke difficulties in tasks such as these. However, had H.P. suffered from any visual perceptual impairment, deficits would have been expected in other tasks necessitating visuospatial processing, such as the Rey–Osterrieth complex figure or Raven's progressive matrices. This was shown not to be the case, his scores on the latter being even above average. A more detailed look at the impaired tasks shows that, once the individual elements (object parts and cubes) are detected, both tests require the capacity to predict an image resulting from the rotation and translation of the elements in space. As was observed, his capacity to rotate objects was impaired, leading to the hypothesis that this particular cognitive operation was deficient. In point of fact, the complete tetrad also appears to require visuospatial processing in the form of translation, rotation or other transformations of visual mental images.

Indeed, writing experiments showed that letters b, q, d and p could be copied correctly, but that the patient could not reproduce them without a model (i.e. to dictation). It is important to note that H.P. had lost his writing skills and each letter had to be reconstructed under visual control. When writing the four letters b, q, d and p, H.P. always began by a circle and only then added the ascending or descending stroke. The four letters share the common characteristic that they are formed by a circle with an adjacent stroke. Their specificity is defined by the direction of rotation of the circle (clockwise or counterclockwise) and the

orientation (up–down) of the stroke. Production of these letters necessitates a spatial organization of the vertical stroke with the circle. Spatial proximity of the letters is probably responsible for up–down confusions, those confusions being also present at the mental imagery level (Zesiger *et al.*, 1997).

The calculation battery showed that the most important impairment concerns digit processing which relies on a visual support, described by Dehaene and Cohen (Dehaene and Cohen, 1997) as a numerical line. According to Dehaene (Dehaene, 1992), numbers are mentally represented in an analogue format, from left-to-right in French writing culture, and are associated with a specific representational space. H.P. had difficulties even in simple tasks such as counting. Although this is generally considered an automatized language function, it has been suggested that the task may be associated with a spatial representation [for example, the so-called SNARC (spatial–numerical association of response codes) effect described by Dehaene (Dehaene, 1992)]. The SNARC effect was evidenced in controls in the following manner. Digits from 0–9 were presented visually and subjects were asked to respond as quickly as possible with a left- or right-hand button-press if the digit was greater or smaller than a middle value (4.5). Responses to smaller values (e.g., 1 or 2) were faster with the left hand than with the right and, conversely, reponses to greater numbers (e.g., 8 or 9) were faster with the right hand than with the left. This is thought to reflect the fact that the subject's representation of digits is spatially mapped. The same opinion is held by Seron (Seron, 1993) who hypothesized that since numbers are learned in a fixed order, they must be represented in a spatial configuration which mirrors their sequence of appearance. Evidence for a learned spatial representation of numbers was further given by Dehaene (Dehaene, 1992) who demonstrated that the SNARC effect was reversed in Iranian subjects, who write from right to left. The dissociation between H.P.'s preserved language capacities and impaired counting abilities suggests that the latter is not related to the production of an automatic language sequence. Rather, the fact that H.P. requires the use of his fingers to count suggests that a visual support is necessary. In line with the SNARC effect, H.P. may have trouble in mentally representing the progression of a cursor along the spatially represented numeric continuum. Moreover, H.P. had more difficulty with perceptual estimation of quantities, which necessitated correct inferences from visual imagery, than with cognitive estimation of quantities.

Finally, finger agnosia was present only when visual support was suppressed, which requires a visual image transformation, and left–right discrimination was more impaired when the patient had to make a mental rotation (incongruous position) as in Gold and colleagues' (Gold *et al.*, 1995) patient.

Consequently, we postulate that the difficulties in mental transformations of images constitute a fundamental deficit for H.P.

Given the fact that H.P. presents a difficulty in mental image transformation in addition to the Gerstmann's syndrome tetrad, the question may be raised if the term 'pure' is really adequate. In other words, is our case really a 'pure' Gerstmann's syndrome in which the common denominator has been extracted, or is it just a case of Gerstmann's syndrome associated with a deficit in the manipulation of mental images?

The concept of purity depends on the extent of the examination, as well as the theoretical and historical background (e.g. it is used with reference to the presence or absence of other deficits considered important at that specific period in time). Generally, the term pure Gerstmann's syndrome is employed when the syndrome is seen in the absence of phasic (namely alexia), praxic (usually constructional apraxia) and verbal or visual memory disturbances (Roeltgen *et al.*, 1983). To that extent, our case study may be considered as a pure Gerstmann's syndrome. However, in our attempt to establish the common psychological denominator of the syndrome, a number of tests were carried out that had never been considered before in this context. It is retrospectively impossible to determine whether other cases of Gerstmann's syndrome described as pure suffered the same visuospatial manipulative disorder. Thus, the question will remain open until a case of pure Gerstmann's syndrome without this disorder is found. Nevertheless, the relevance of the concept of visuospatial image transformation in explaining the particularities of all four elements of our Gerstmann's syndrome case leads us to hypothesize that it is indeed the common denominator of the syndrome. Thus, we posit that pure Gerstmann's syndrome without such a disorder will never be found.

Concerning the anatomical data, MRI analysis shows a discrete subcortical parietal inferior stroke, located deep in the inferior part of the gyrus angularis and the superior posterior part of T1 and impairing the myelinated fibres afferent to these cortical areas. According to animal studies, the inferior parietal lobe and the superior-posterior region of T1 are part of the occipitodorsal visual system and contain cells which respond to direction, rotation, and size changes of visual stimuli (Andersen, 1987). Electrophysiological and tracing studies in animals have shown that these cortical regions, called areas 7a, have strong communications with other associative visual areas, like V2, V3 and area MT, involved in integrating information concerning movement (Zeki, 1991). According to Levine and colleagues (Levine *et al.*, 1985), the same neuronal networks involved in such visuospatial skills are also implicated in visuospatial imagery. Along the same line, we hypothesize that networks necessary to process visual motion and mental transformations of images are identical. In H.P., such subcortical lesion situated deep in the angular

gyrus can be a source of disconnection between motion sensitive fibres and the angular gyrus.

The second MRI, carried out 3 years after the stroke, also showed a late degeneration of the posterior callosal fibres, which suggests an interruption of the interhemispheric pathways. This finding points to the fact that there must also be an interparietal disconnection. Such a dysconnection has been suggested in one case of Gerstmann's syndrome to be due to diffuse subcortical HIV-1 induced encephalopathy (Cirelli *et al.*, 1994). However the extent of the lesions made the interpretation of this finding difficult. Recent PET studies dealing with the role of the parietal lobes suggest a potential importance of interhemispheric dysconnection in Gerstmann's syndrome. Mental multiplication and number comparisons activated respectively the inferior parietal regions and the superior temporal regions on both sides (Dehaene *et al.*, 1996). Moreover, execution of overlearned graphomotor writing pattern induces a bilateral posterior parietal cortical activation (Seitz *et al.*, 1997). Bonda and colleagues (Bonda *et al.*, 1995) showed that the recognition of picture of left versus right hands produces activation of the superior and inferior parietal areas. They postulated that such tasks are in fact mental transformations of one's own hand position and that the neural substrates of these mental transformations are located in the dorsomedial visual system. These three studies show separately the importance of the bilateral inferior parietal areas in the different cognitive activities specifically impaired in Gerstmann's syndrome. Furthermore, other imagery studies during mental transformation tasks (Cohen *et al.*, 1996; Alivisatos and Petrides, 1997; Pegna *et al.*, 1997) underline the importance of parietal activation in manipulation of mental visuospatial imagery.

In conclusion, our behavioural data suggests that a difficulty in mental manipulation of images is a fundamental deficit in Gerstmann's syndrome. The anatomical data show a disruption of the left occipitodorsal system and an interhemispheric dysconnection involving the homotopical parietal areas. These anatomical clinical data suggests that the integrity of this bilateral dorsomedial network is necessary in order to allow the realization of any task requiring the manipulation of mental images on which Gerstmann's syndrome depends.

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