WHAT THE EYES PERCEIVE, THE BRAIN IGNORES: A CASE OF PURE UNILATERAL REPRESENTATIONAL NEGLECT*

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ABSTRACT

Bisiach and Luzzatti (1978) provided evidence that unilateral spatial neglect is not only a disorder of visual perception, but also can affect mental representations such that patients fail to report the left side of scenes or objects in mental imagery. However in case reports of representational neglect generally it is accompanied by perceptual neglect.

We report a rare occurrence of a patient (NL) who presents a persistent unilateral neglect which appears to be limited to visual imagery. The deficit appeared in tasks which require the formation and manipulation of new visuo-spatial representations as well as those which require access to information about familiar scenes. The patient, who had a lesion in the right parietal lobe, showed no evidence of perceptual or personal neglect, although there was some evidence of visual extinction.

We argue that the concept of visuo-spatial working memory can provide a framework within which to interpret aspects of the representational form of neglect, whether or not it is accompanied by perceptual neglect.

INTRODUCTION

Unilateral spatial neglect (USN) generally is defined as an impaired ability to detect or respond to stimuli in spatial locations contralateral to the damaged cerebral hemisphere in the absence of peripheral deficits. It is more frequent, severe, and long-lasting after a right parietal lobe lesion (Vallar, 1993). This definition of USN has been based primarily on observed deficits in visual-perceptual tasks. However it has become clear that USN is not an impairment limited to the processes of visual perception (Zingerle, 1913) but also can compromise the "left side" of visual mental representations, sometimes referred to as visual mental images (De Renzi, Faglioni and Scotti, 1970).

Two of the best known cases of representational neglect were described by Bisiach and Luzzatti (1978). They tested two patients showing left USN, asking them to describe from memory a very familiar piazza, the Cathedral Square (Piazza del Duomo) in Milan. The patients had first to give their description from a given imagined viewpoint at one end of the piazza, and then to describe the same piazza from the opposite end. From both viewpoints they accurately

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described only the buildings on the right side of their mental image, failing to mention those on the left side. This study gave rise to the suggestion that USN also may involve mental representations in the absence of physical stimulation. This impairment was thought to arise from damage to the brain hemisphere opposite to the side of the neglected information in the mental representation. In this sense, the representation was functioning like the visual perceptual environment. Subsequent studies have supported the idea that USN involves mental representations (Bisiach, Luzzatti and Perani, 1979; Bisiach, Capitani, Luzzatti et al., 1981; Baxter and Warrington, 1983; Ogden, 1985; Barbut and Gazzaniga, 1987; Caramazza and Hillis, 1990a, 1990b; Halligan and Marshall, 1992) and it is now agreed that disorders in the perceptual and the representational domain can occur together (Grossi, Modafferi, Pelosi et al., 1989; Rizzolatti and Berti, 1993; Bartolomeo, D'Erme and Gainotti, 1994).

The interpretation of these findings is still very much a matter for debate. USN has been attributed to an impairment within a system that usually modulates the ability to select and orient to stimuli in the visual world (Popperlreuter, 1923). It has also been attributed to an impairment in orienting attention to the contralesional side (Riddoch and Humphreys, 1983); or to a strong tendency to orient attention to the ipsilesional side (Kinsbourne, 1970); or to the impaired disengagement of attention from stimuli located in the ipsilesional side (Posner, Walker, Friedrich et al., 1987). However these attentional theories do not incorporate the nature of the cognitive system that might be responsible for the mental representation. Nor do these theories adequately address how such a cognitive system might support covert attention to different parts of the representation.

What kind of theoretical model might account for the representational form of neglect? Clearly one approach to resolving this issue is to explore theories of mental representation in the healthy brain and investigate whether any such theory can offer an account of representational deficits. One candidate theory arises from the mooted close relationship between processes involved in mental representation and those involved in working memory. Working memory refers to a set of cognitive mechanisms thought to be responsible for online processing and temporary storage of information. In one widely adopted model, working memory comprises an executive coordinator and two systems specialised for dealing respectively with visuo-spatial information and with verbal information (Baddeley and Hitch, 1974; Baddeley, 1986, 1996). This model has been singularly successful in accounting for a wide range of cognitive deficits, although its use in accounting for specifically visuo-spatial deficits has thus far been rather limited (for recent reviews see Della Sala and Logie, 1993; Logie, 1995).

Baddeley and Lieberman (1980) suggested that patients such as those described by Bisiach and Luzzatti "...have a defect which is associated with spatial working memory" (p. 23) and, using the analogy of the specialised visuo-spatial system as a screen on which spatial information may be represented, they also stated that "...one might conclude either that part of the screen was inoperative or that the process of scanning the screen was defective..." (p. 23). The view has been supported by other authors (e.g. Sunderland, 1984; Shallice, 1988), and Bisiach (1993, p. 439) has endorsed the suggestion of a partially inoperative visuo-spatial component of working memory. However as yet the hypothesis has not been investigated in any detail. Moreover, in the vast majority of cases of representational neglect that have been reported, there has been concomitant visual perceptual neglect. Therefore it is not clear whether the impairment arises primarily from impoverished perceptual input on the neglected side or from an impaired representational system. One way to explore this further would be if patients were found with a representational impairment but with intact visual perception.

Guariglia, Padovani, Pantano et al. (1993) described one such patient who showed a persistent disorder in visual imagery for familiar piazzas in the absence of perceptual USN. During the description of familiar places from different vantage points his performance was very similar to Bisiach and Luzzatti's patients. However on perceptually based tests he appeared to perform normally. Subsequently, Bartolomeo et al. (1994) analysed the relationship between visuo-spatial and representational USN in 30 right- and 30 left-brain-damaged patients and they found no cases of representational neglect in the absence of perceptual deficits. In a follow-up eight months after the stroke one of their patients, with a right hemisphere lesions, showed neglect limited to mental representations, and the signs of visual perceptual neglect were no longer present. These authors suggested that different compensatory mechanisms rather than lesions in different components of the cognitive architecture were responsible for this result. Bartolomeo and colleagues noted that the same could be true for the Guariglia et al. patient who was tested 16 months after the stroke. Notwithstanding this observation, the patient reported by Guariglia et al. suggests that the dissociation is possible. In this paper we pursue this possibility by reporting the case of a patient (NL) who, following an ischaemic lesion in the right anterior parietal lobe, presented a persistent unilateral neglect limited to visuo-spatial imagery. We examined the patient for a period of several weeks after the stroke, and subsequently. Following the formal description of the patient, we shall discuss the pattern of data obtained in the context of the visuo-spatial working memory hypothesis referred to above.

Case Report

NL, a 67 year old right-handed man with eight years of education, was admitted on December 28, 1993 to a Neurological Unit after an ischaemic stroke which caused a left hemiparesis and left hemianaesthesia. A Computerised Tomography scan (13/01/1994) showed a large area of hypodensity in the right parietal lobe. Two months after the vascular accident he was co-operative and was oriented in time and in space. His speech was fluent and he did not show difficulties referable to aphasic deficits. He could easily recognise familiar faces, showing no evidence of prosopagnosia.

Although no formal psychometric testing for perceptual neglect took place immediately after the stroke there are a number of sources of evidence to suggest that perceptual neglect was not present even at that stage. Doctors and nurses did not notice any typical neglect behaviours: he was reported to perceive stimuli coming from each side of the visual field and successfully to reach out to targets in both the right and left hemifields. Also his wife was asked to complete a questionnaire concerning several behavioural aspects of neglect, and her responses all were negative. He had a paresis in his left limbs, for which he was given a programme of motor rehabilitation which commenced one week before formal testing.

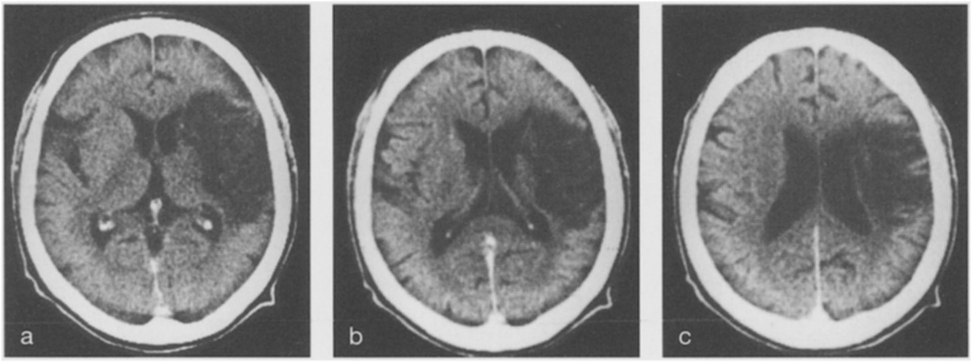

Fig. 1 – *CT Scan of patient NL, which shows an ischemic lesion in the right parietal lobe.*

He was aware of his motor problems, indicating the absence of any anosognosia. Relatives reported that NL was depressed and often cried.

Two and a half months after the stroke, he was sent to the first two authors for a routine neuropsychological assessment. A new CT scan (22/03/1994) confirmed the previous findings (see Figure 1).

General Neuropsychological Examination

NL's scores on a variety of neuropsychological tests are shown in Table I. His performance on the Mini Mental State Examination (Folstein, Folstein and McHugh, 1975) was adequate for his socio-educational background (Measso,

TABLE I

NL's General Neuropsychological Examination Scores adjusted for age, education and, whenever appropriate, sex (Score range is indicated in parentheses. For details of tests see text).

Test (score range)	NL's scores adjusted for age and education	Controls' inferential 5th centile or cut-off[a]
– Global assessment		
Mini Mental State Examination (0-30)	28.53	23.80
– Short term memory		
Bisyllabic word span (0-10)	4	2.75
Corsi block-tapping span (0-10)	4.25	3.50
– Long Term Memory		
Learning three words lists (0-30)	18.5	7.50
Corsi block-tapping spatial supraspan (0-29.16)	18.86	5.50
Maze learning (15-1)[bc]	12*	6.2[a]
– Intellectual functioning		
Ravens Progressive Matrices (0-36)	22.50	14.75
Verbal abstract reasoning (0-60)	60	32
– Visual Perception		
Unfamiliar face recognition (0-54)	38	34
– Spatial cognition		
Constructional apraxia (0-14)	10.75	7.75
Rey complex figure A (0-36)	25.63*	27.99
Rey complex figure B (0-11) (elements plus details)	11	—
– Acalculia		
Acalculia test (0-21)[c]	21	—

[a] Cut-off is the score two standard deviations below the mean score of the control group.
[b] Error score as trials to criterion.
[c] Not adjusted for age and education.
* Score below the cut-off.

Cavarzeran, Zappalà et al., 1993). He had no verbal or spatial short or long term memory deficits (bisyllabic words span – Baddeley, Thomson and Buchanan, 1975b; learning three words lists test – Novelli, Papagno, Capitani et al., 1986; Corsi's block-tapping test – Milner, 1971; and Corsi's block-tapping supraspan test following the procedure described by Spinnler and Tognoni, 1987). NL was also given a maze learning test (Milner, 1971) which consists of a long pathway through a 10 × 10 grid of buttons. As can be seen in Table I, the number of trials he required to reach criterion performance was higher than that of 5 normal control subjects, but his pattern of errors showed no suggestion of asymmetry. Visuo-spatial and verbal intellectual functions, assessed with Raven's Coloured Progressive Matrices (Basso, Capitani and Laiacona, 1987) and verbal abstract reasoning (Spinnler and Tognoni, 1987) were unimpaired. NL had a normal score on a test for recognition of unfamiliar faces without involving memory (test of facial recognition – Benton and Van Allen, 1968). On two tests for figure copying (constructional apraxia test – Arrigoni and De Renzi, 1964, and the Rey complex figure A – Rey, 1942, 1959) he did not show constructional apraxia nor was there evidence of neglect, but he copied two pictures (one was the Rey figure) rotated 90° anticlockwise (Turnbull, Beschin and Della Sala, 1996). Although scores were independent of the orientation of the pictures, on Rey's figure copy test NL's score fell below the cut-off for normal performance (see Table I). The rotation phenomena could have been the main reason for his poor performance on this copying task but the important point is that there was no evidence of neglect on these tasks. He performed flawlessly on a test assessing mathematical abilities.

In summary NL showed no mental global deterioration, he had no verbal memory deficits, no abstract reasoning difficulties, or signs of acalculia and no deficits in recognizing unfamiliar faces. Nor were there signs of neglect in copying geometrical and abstract patterns. He performed normally on all spatial tasks when the stimuli were present. His only observed spatial impairment was in learning a long pathway through a matrix. This pathway test has been found to be particularly sensitive to spatial disorientation in daily life (Hécaen, Tzortzis and Rondot, 1980).

Assessment of Unilateral Spatial Neglect

Personal Neglect

Personal neglect was assessed with four tests that required movements towards both sides of the body. NL's scores are shown in Table II. Normal scores in the Table are those originally reported by the authors of the tests.

Using the Bisiach, Vallar, Perani et al. (1986) one item test of personal neglect the patient was blindfolded and was required to touch his left hand with his right hand. This test has a score range of 0 to 3 depending on the speed and accuracy of performance. NL promptly reached for the target (score 0 = no personal neglect).

The Fluff test (Beschin and Robertson, in preparation) required the subject to reach for targets in the form of 2 cm circles attached to the front of clothes that he was wearing. On the torso there were three stickers on the left and three on the right of the central midline area, there were six stickers along the patient's left arm, six along the right leg and six along the left leg. No targets were placed on the right arm because the task was performed with this arm. In total there were 15 targets on the left side of the body and nine targets on the right. The patient was blindfold when the targets were attached and was not informed as to how many targets were present. In the first condition he had to remove all the targets while blindfold. In the second condition the blindfold was removed. Results are shown in Table II from which it is clear that NL successfully reached all the targets in the "no blindfold" condition, but that in the "blindfold" condition he missed 12 of the 15 targets on his left side and performed flawlessly with the targets on his right. Because of his poor performance in the blindfold condition, this condition was repeated the following day and very similar results were obtained (see Table II).

On the Comb test and Razor test (Beschin and Robertson, in preparation) NL was asked to simulate respectively combing his hair and shaving himself for 30 seconds. The tasks were performed twice with an intervening delay of 15 days. Results are shown in Table II presented as the difference in the number of strokes on the right and left side of the body. NL's scores were compared with norms collected from 64 brain damaged and normal subjects (Beschin and Robertson, in preparation. His performance on the test was above the cut-off score on the first test, but below the cut-off on the second test.

In summary NL performed normally on tasks that permitted visual input and feedback. He showed personal neglect only on two tasks. In one of these he was blindfold and therefore would have to rely on a mental representation of his body. In the Comb and Razor test he scored just below the cut-off on retest. In the case of this test, there is only partial visual input as to the position of his arm, and he would be unable visually to monitor the parts of his body involved (hair and chin). The fact that he could watch his arm movements but did not have complete visual information may have led to his borderline performance on this task which relies at least in part on a mental representation of his face and head.

TABLE II

NL's Performance on Tasks Assessing Personal Neglect (Score range is indicated in parentheses. For details of tests see text)

Tests (score range)		NL's scores		Cut-off	
		Left	Right	Left	Right
Personal neglect test (3-0)		0 (no defect)		—	
Fluff test – eyes open (left 0-15; right 0-9)		15	9	15	9
Fluff test – eyes closed (left 0-15; right 0-9)	test	3*	9	13.5	9
	retest	0*	9		
Comb and Razor test (0-inf.)	test	36.6%		34.6%[a]	
	retest	29.9%*			

Cut-off is the score two standard deviations below the mean score of the control group.
+ Score below cut-off.
[a] The score is expressed as a percentage of strokes on the left relative to the total number of strokes on the left, centre, and right.

Perceptual Neglect

NL was given tests to assess the presence of visual perceptual neglect. The patient's scores are shown in Table III. NL performed at ceiling on the Stars Cancellation Test (Wilson, Cockburn and Halligan, 1987), Lines Cancellation Test (Albert, 1973), Wundt-Jastrow Area Illusion Test (Massironi, Antonucci, Pizzamiglio et al., 1988) and Perceptive Discrimination Between Objects Test (Bisiach and Rusconi, 1990). On the Bells Test (Gauthier, Dehaut and Joanette, 1989) he missed only one target on the left side the first time, and one target on the right side the second time, and with this test presence of neglect is assumed only with three or more omissions on one side (Vallar, Sterzi, Bottini et al., 1990). On the Letter Cancellation Test (Diller and Weinberg, 1977) his performance was very good. He missed one target on the left side and the presence of neglect in this task is assumed only if there are four or more omissions on one side only (Zoccolotti, Antonucci, Judica et al., 1989). NL's scores on two horizontal lines bisection tests and a vertical lines bisection test were compared with those of five controls and no difference emerged. Moreover, NL's performance on these tests showed no difference between the right and left side. NL also was tested on four separate tests using verbal stimuli (see Table III). The tests comprised the Reading Horizontal Sentences Test (De Renzi and Spinnler, 1966; Zoccolotti et al., 1989), the Reading Vertical Sentences Test (Zoccolotti et al., 1989), and two single word reading tests devised for this study, one involving normal words and the other involving mirror reversed words. In these last two tests a list of thirty words of varying frequency, concreteness and length (4-8 letters) were printed in large capital letters in normal

TABLE III

NL's Performance on Tasks Assessing Perceptual Neglect (Score range is indicated in parentheses. For details of tests see text)

Test (score range)	NL's scores	Cut-off range
Star cancellation (0-54)	54 (L−R=0)	L−R≥±2
Lines cancellation (0-40) test	40 (L−R=0)	L−R≥±1
retest	40 (L−R=0)	
Wundt-Jastrow area illusion (0-40)	40	—
Perceptive discrimination between objects (0-48)	48	—
Bells test (0-35) test	34 (L−R=−1)	L−R≥±3
retest	34 (L−R=+1)	
Letter cancellation (0-104)	103 (L−R=−1)	L−R≥±4
Horizontal lines bisection[a] (9 single lines) (−720/+720 mm)	+0.17	+6/−5.6
Horizontal lines bisection[a] (5 multiple lines sets) (−1060/+1060 mm)	+0.11	+2.8/−3.9
Vertical lines bisection[a] (5 single lines) (−460/+460 mm)	+0.28	+5.1/−8.7
Word reading (0-30)	30	—
Mirror word reading (0-30)	30	—
Reading horizontal sentences (0-6)	6	—
Reading vertical sentences (0-2)	2	—

Cut-off is the score two standard deviations below the mean score of the control group. L=left; R=right.
[a] The distance from midpoint is expressed in millimetres. A minus sign indicates an error towards the left or towards the bottom. A plus indicates errors towards the right or the top.

orientation or mirror reversed and presented one at a time for the subject to read aloud. On all four tests, NL performed at ceiling (see Table III)

Extinction

Visual extinction was assessed with a newly devised automated test (visual extinction test). NL was asked to fixate a 1.5 cm × 1.5 cm cross displayed in the centre of the computer screen. His task was to detect single or double targets comprising 5 mm white dots displayed for 100 ms. In the case of single targets these appeared either about five visual degrees to the left or to the right. In the case of double targets, these involved either one single target on each side, or two dots presented five degrees apart either on the left or on the right. When the two dots were presented on the same side, they could be presented one above the other or side by side, and in this last case the target nearer to the centre of the screen was about 2.5 visual degrees from the point of fixation. The order of presentation was randomised. The patient had to decide if one or two targets had been displayed and then to indicate on which side of the screen they had appeared. He indicated his decision by pressing one of two different response-keys with his right hand.

Results are shown in Table IV, from which it is clear that he had difficulty only with left sided targets when they were presented simultaneously with one target on the right.

To assess the presence of auditory neglect and of auditory extinction, NL was presented with sounds via headphones. Ten sounds were presented to the left ear, ten sounds to the right, and ten sounds to both ears simultaneously. The order of presentation was random and the sound was always the same recorded repetitively. Table IV shows that NL had very little difficulty with this task, although on five occasions he reported that he had heard the stimulus in both ears when it had only been presented on the right, and in the double stimulation condition he missed 20% of left stimuli. With few exceptions (e.g. De Renzi, Gentilini and Pattacini, 1984), this level of performance usually is

TABLE IV

NL's Performance on Tasks Assessing Extinction (Score range is given in parentheses)

Test (score range)		NL's scores	
		Left	Right
Visual extinction			
Single stimulus presentation	(0-10 each side)	10 (100%)	9 (90%)
Double unilateral presentation			
– Vertical	(0-12 dots each side)	11 (92%)	11 (92%)
– Horizontal	(0-12 dots each side)	12 (100%)	12 (100%)
Double bilateral presentation	(0-8 each side)	0 (0%)	8 (100%)
Auditory extinction			
Single stimulus presentation	(0-10 each side)	10 (100%)	10 (100%)
Double bilateral presentation	(0-10 each side)	8 (80%)	10 (100%)
Tactile extinction			
Single stimulus presentation	(0-10 each side)	10 (100%)	10 (100%)
Double unilateral presentation	(0-10 each side)	10 (100%)	10 (100%)

not considered to be evidence of extinction (see e.g. Bisiach et al., 1986; Grote, Pierre-Louis, Smith et al., 1995; Arboix, Junqué, Vendrell et al., 1990).

We also assessed tactile extinction, but only with his right hand because of the sensory problems in his left hand (hemianaesthesia). NL was given ten stimuli on the left side of the back of the hand, ten on the right side and ten on both sides following a procedure used by Moscovitch and Behrmann (1994). NL's performance was flawless in all conditions.

In summary, NL showed neither a disorder of perceptual neglect nor of directional hypometria. In fact he performed normally on all of the tests requiring movements of the arm on the left side. He showed no asymmetry in perceiving targets in upper or lower space. It appears that he did show some evidence of visual extinction but did not show auditory of tactile extinction, although tactile extinction cannot be completely ruled out as we were unable to test his left hand. It is worth emphasising that extinction was clearly dissociated from visuo-spatial neglect. This finding of visual extinction in the absence of visuo-spatial neglect adds to other similar cases which can be gleaned from the literature (Vallar and Perani, 1986; Barbieri and De Renzi, 1989; Vallar, Rusconi and Bisiach, 1994a; Di Pellegrino and De Renzi, 1995). This distinction coupled with reports of the converse, namely visuo-spatial neglect without extinction (Kartsounis and Findley, 1994), supports the notion that the two phenomena have to be considered symptoms of two different processes.

Representational Neglect

Representations from Long-term Memory. Data for the tests of representational neglect are shown in Table V. NL was given a spelling test in which he was presented aurally with 80 words and 10 non-words, and had to respond by spelling each word or non-word aloud. Thirty of the words were

TABLE V

NL's Performance on Tasks Assessing Representational Neglect (Score range is indicated in parentheses)

Test (score range)	NL's scores		Cut-off	
Mirror reversed reading (0-30)	30		—	
Spelling test (0-90 word error score)	24 (L=11; R=9; mid=4)		—	
Letters in 24 words spelled incorrectly				
Left (0-75 letter error score)	23		—	
Right (0-75 letter error score)	13		—	
Centre (0-11 letter error score)	6		—	
Verbal description of complex scene (Scores Left 0-22, Right 0-18)				
Immediate recall	L=5	R=5	L=5.0	R=3.18
Recall delay of 15 minutes	L=1*	R=4	L=4.0	R=2.28
Recall delay of 2 days	L=2*	R=4	L=4.5	R=0.28
Tactile maze				
Eyes closed (0-48) test	32*		45.31	
retest	29*			
Eyes open (0-49)	48			

Cut-off is the score two standard deviations below the mean score of the control group.
L=left; R=right; mid=middle of the word.
* Score below cut-off.

chosen to have transparent sound to spelling correspondence in Italian (e.g. c-a-s-a). The remaining words and non words had no such simple correspondence (e.g. scienza – the initial sc is pronounced as "sh" in English). From Table V it is clear that the patient made errors on 24 of the items. Within those 24 items, Table V also shows the number of letters incorrect according to whether they would have been on the left, on the right or in the middle of the words, had they been printed or represented in a visual mental image. Although there are no control data for this test, more errors were derived from the left of the words.

NL showed clear differences between the left side and the right side of mental images of very familiar piazzas (Bisiach and Luzzatti, 1978; Guariglia et al., 1993) and geographical areas (Halligan and Marshall, 1992; Bartolomeo et al., 1994). In one form of the task, NL was asked to mentally visualise a piazza (see Figure 2) in the town where he lives and with which he is very familiar. In a second version of the task he was asked to imagine himself in Sardegna looking towards Roma or vice versa (see Figure 3). When he had to describe them from a given vantage point (Figures 2a and 3a), he consistently reported several elements on the right side (8 details for the image of the piazza and 4 details for the geographical locations) and very few on the left side (2 details for the piazza and 9 details for the geographical image). Ten days after this first test session the patient was asked to describe the same mental images from the opposite perspective (Figures 2b and 3b), he again reported more elements on the right (9 details for the piazza and 3 details for geographical locations) than on the left (2 details for the piazza and 1 detail for the geographical locations). The ten day delay between changes of viewpoint was introduced to avoid the possibility that he used verbal labels for some of the details when changing between vantage points (Logie, 1995). On these tasks the patient showed some characteristics of neglect behaviour. Specifically he always started to describe the images from the right side, he was much more accurate and he spent more time reporting the details from the right of the imagined scene. On two occasions he reported details that were on the right as being on the left or

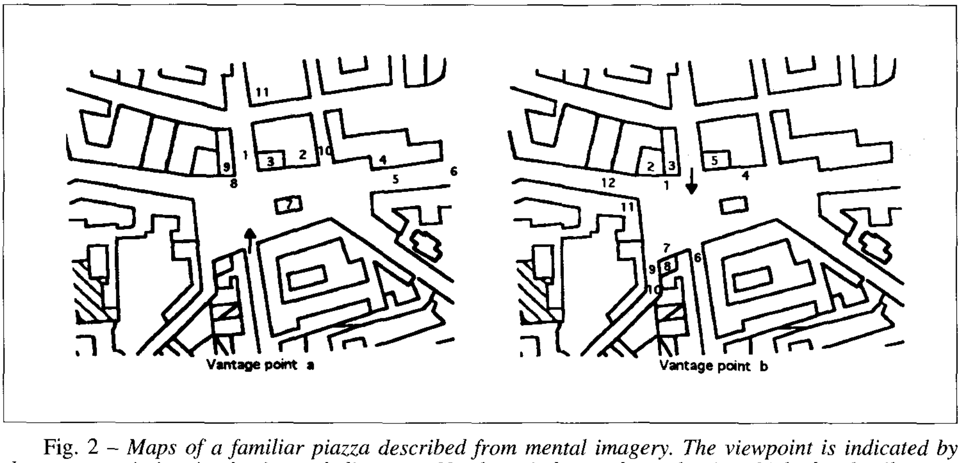

Fig. 2 – *Maps of a familiar piazza described from mental imagery. The viewpoint is indicated by the arrow pointing in the imaged direction. Numbers indicate the order in which the details were reported.*

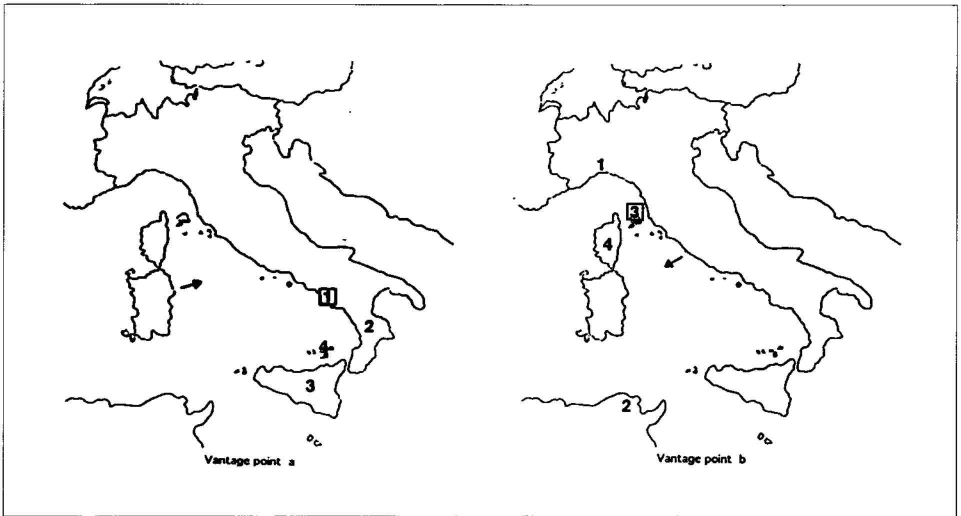

Fig. 3 – *Maps of Italy described from mental imagery. The viewpoint, from the Sardegna coast (a) and from Roma (b), is indicated by the arrow pointing in the imaged direction. Numbers indicate the order in which the details have been reported. Numbers surrounded by a square refer to the details transferred from the right to the left side.*

in the centre. For example when describing his image from Sardegna he said "... on the left of Roma, towards the North, there is Napoli...". The city of Napoli is actually south of Roma (to the right from Sardegna), and this would be familiar to most Italians.

Another interesting aspect of NL's visual imagery ability comes from drawings from memory. He was asked to use a pencil to draw each of the following: A tractor, a house, a daisy, a watch, a telephone, a person, a horse, a bear, a table, a car, a cat, and a map of Italy. Using coloured pencils he also spontaneously drew a landscape with which he was familiar. In all cases there were striking differences in the level of detail depicted on the left and right. The right side of his drawings were very detailed, but on the left he missed salient elements and those details which were present tended to be drawn in the wrong place (see Figure 4a). After 20 days NL was shown some of his drawings and was asked to describe them. He recognised that they had been drawn by him and he identified omissions and erroneous spatial relationships among elements of the drawings. One notable example was his description of the drawing of the landscape that he used to see when he was driving toward his holiday house (Figure 4b): he noticed that the picture was unbalanced on the right side of the sheet but when he was asked the reason for this, he answered: "... the left of the mountain is all grey because it is all rock...". One possible alternative interpretation is that the difficulty shown in these tasks arose from a sensory motor processing difficulty.

However were this the case, we would have expected the difficulty also to show up in the copying tasks described earlier. Therefore the impairment seems to arise when NL has to rely on information in memory.

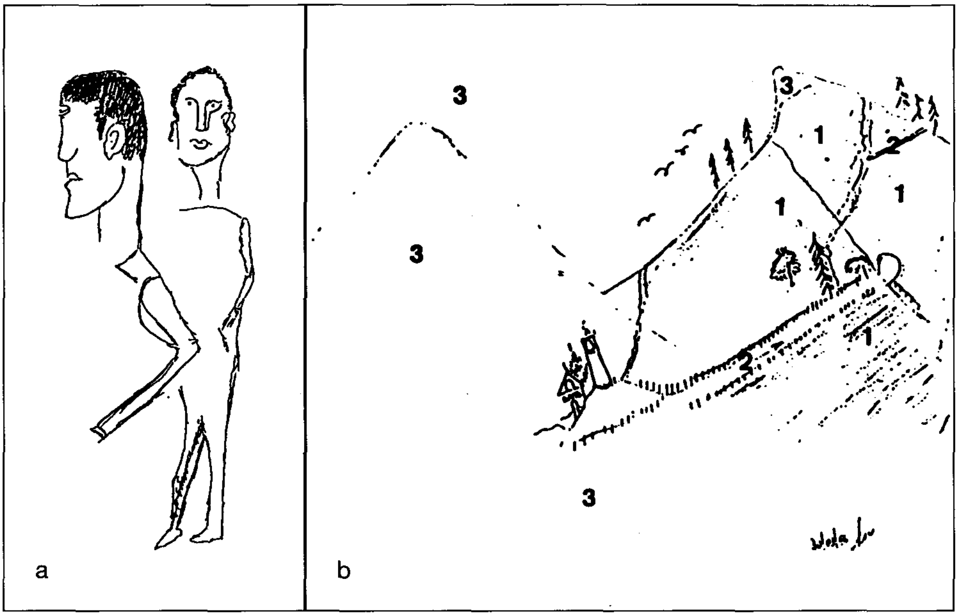

Fig. 4 – *Drawings from memory of a person (a) and of a familiar landscape (b). In picture a, the patient drew two ears on right side. Picture b was originally in colour, and in this reproduction numbers indicate the colours that the patient used: 1 = green; 2 = brown; 3 = blank.*

Representations Based on Novel Material. The pattern described thus far suggests that NL showed evidence of neglect when performing representational tasks based on mental images processed and stored in long term memory before the lesion. The next obvious question is how he coped with representational tasks that required the generation of new mental representations of stimuli to which he had been exposed only in the testing sessions and he had not seen before. NL was asked to remember objects, people and animals shown in a complex picture which depicted a farm scene. The picture had 22 discrete items on the left and 18 on the right. He examined the picture in silence for 2 minutes after which the picture was covered and he was asked for immediate spoken recall of the items in the picture. Items were scored correctly only if he recalled a complete item (e.g. a tree, a tractor, a hen etc.). After recalling as many items as possible, he was asked to produce a drawing of the picture from memory, using a blank sheet identical in size to the original scene (25 × 24 cm). Results are reported in Table V. The number of items reported orally was the same from both sides of the picture but in his drawing from memory the spatial relations were incorrect among the details from the left of the picture. Subsequent spoken recalls were required 15 minutes and two days later. In contrast to immediate recall, on both occasions he recalled fewer details from the left of the picture than from the right (see Table V). It is clear from the cut off scores shown in Table V that, if anything, the controls recalled slightly more items from the left than from the right of the picture.

NL's ability to create and manipulate mental representations of new information also was assessed by a tactile task, the Tactile Exploration Maze Test (Beschin, Cazzani, Cubelli et al., 1996). This test has been thought to rely on spatial representation and not visual perception (De Renzi, Faglioni and Scotti, 1970). The test involves a 33 cm square maze cut into a 40 cm by 50 cm wooden board (see Figure 5c). Four alleys (2 cm wide and 3 mm deep) spread from the centre of the maze branching into eight lateral arms, each of which ends in a 10.5 cm segment. At the extreme ends of each segment there is a hollow designed to hold a 2.4 cm marble which is used as a target. Each target position is therefore at the same distance from the centre. NL had his forefinger in the centre of the maze and he was asked to move it along the alleys of the maze. His task was to find as quickly as possible a marble placed in one of the extremities of the maze. The whole test consisted of 48 randomised trials (three for each of the 16 possible positions). The time limit for each trial was 120 sec and any failure to locate the target within the time limit was scored as an omission. A frequency of omissions on the left side greater than 3 times that on the right was considered evidence of neglect (Beschin et al., 1996). The test was given three times. On the first two occasions (which were one month apart), visual input was prevented (the patient was blindfold). During the first test NL missed the marble 16/48 times, 3 times when it was on the right side and 13 times when it was on the left. On the second occasion NL missed the marble 19/48 times, 17 times when it was on the left side and two on the right. Normal subjects are virtually at ceiling on this task (Beschin et al., 1996). At the end of this session NL was required to draw what he thought the unseen maze looked like. NL's drawing of the maze is shown in Figure 5a, along with a normal subject's typical reproduction (5b) and the real maze configuration (5c). NL's representation is completely devoid of details from the left side of the maze. On the third test session, with eyes open and uncovered, NL found all the marbles (48/48) without difficulty. In this condition he took longer to search on the right (mean = 4.2 s) than on the left (mean = 3.3 s). However the bottom right corner of the maze was occluded by his right elbow during much of the task, and when the data based on this part of the maze were excluded, there was no difference in the mean search times for left and right ($F = 3.85$; d.f. = 1, 38; ns). In either case, there is no hint of a left neglect in his searching time with eyes open.

In summary, NL showed no evidence of perceptual neglect but he did show a unilateral neglect restricted to visual imagery similar to the pattern reported by Guariglia et al. (1993). However unlike the patient described by Guariglia and colleagues, NL also showed a representational neglect in tasks that require the creation and manipulation of new mental representations.

Brooks Matrix

As mentioned in the introduction, one focus of this paper is to explore the feasibility of the visuo-spatial working memory model hypothesis in the context of representational neglect. One of the tasks most widely used in the visuo-spatial working memory literature (Baddeley, Grant, Wight et al., 1975a; Baddeley and Lieberman, 1980; Farah, Hammond, Levine et al., 1988; Hanley

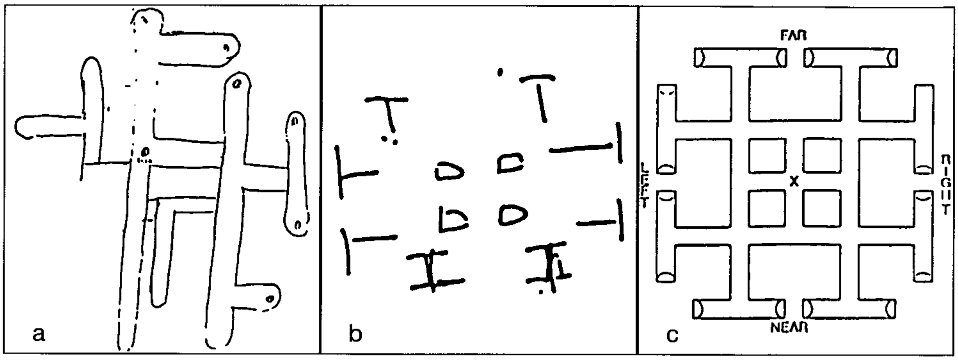

Fig. 5 – *Graphical reproduction of tactile maze. (a) NL's reproduction, (b) performance of a typical normal subject, (c) configuration of the real maze in which the x indicates the starting point.*

and Pearson, 1990; Hanley, Young and Pearson, 1991) was originally devised by Brooks (1967). A spatial version of this task has been shown to rely heavily on visuo-spatial working memory, while a more verbally oriented version relies on the verbal working memory system. In particular, performance of the spatial version of the task has been shown to be disrupted by concurrent overt movement such as tracking (Baddeley et al., 1975a), or by tapping out a pattern on a table (e.g. Quinn and Ralston, 1986). This pattern of data has been taken to support the view that the production of overt movement and the encoding and recall of imagined spatial patterns rely on visuo-spatial working memory (for a review see Logie, 1995).

In the Brooks' task, the subject is asked to imagine placing consecutive numbers in consecutive squares of a four by four matrix, starting from the second column on the second row. In one (spatial) version of the task, NL listened to the instructions as to the sequence of directions around the matrix, moving from square to square (e.g. up-2, right-3, down-4). Once the presented sequence was completed the patient responded with verbatim vocal recall of the series of directions and numbers. Next, a complementary (verbal) form was used where the patient again had to retain a sequence of sentences, except that the words "up, down, left and right" were replaced by the words "good, bad, slow and quick". This resulted in a set of nonsense items (e.g. good-2, slow-3, bad-4), which were to be retained without the use of any form of visual image, perhaps by subvocal rehearsal. In both conditions, NL was presented with sequences of increasing length (range 2-9) with 3 trials for each length of sequence and the score (span) was the maximum length for which NL correctly recalled at least two out of three sequences.

NL's scores are shown in Table VI together with those of the control group matched for age. His performance was in the normal range in the verbal task, but when performing the spatial task he obtained a score which was two standard deviations below the mean for the controls.

To investigate if these results arose because of heavy reliance on visuo-spatial memory in the spatial version of the task, NL was again given the spatial task but with the matrix present. This then provided perceptual information (stimulus

TABLE VI
NL's Performance on the Brooks Matrix Test (Score range is indicated in parentheses)

Conditions (score range)	NL's scores	Cut-off	Controls' mean	Controls' standard deviation
Verbal span (0-9)	3	2.40	3.2	0.4
Spatial span (0-9)	2*	2.46	4.8	1.2
Spatial span with stimulus support (0-9)	8	2.48	6.4	1.9
"No-memory" perceptual task (0-9)	8	7.62	8.6	0.5
"No-memory" imagery task (0-9)	2*	5.00	6.6	0.8

Cut-off is the score two standard deviations below the mean score of a control group.
* Score below cut-off.

support) about the form of the matrix, thereby reducing the load on the visuo-spatial memory system. Under these conditions, NL's performance improved dramatically (see Table VI). Next NL was tested with two further tasks in which he was not required to retain the sequence of directions (no memory load). NL was given nine sequences and each sequence continued until the patient reported that the path described by the sequence of directions led off the edge of the matrix. In one case he could see the matrix during the task ("no memory perceptual task"), and his performance did not differ from the mean of the control subjects (see Table VI). In the other case there was no stimulus support provided and the patient had to form and retain a mental image of the matrix ("no-memory imagery task"). His scores in this condition fell two standard deviations below the mean of the control group.

In conclusion, NL demonstrated that he was able to perform spatial "scan" operations when he was provided with external support via visual perception and did not have to rely heavily on visual imagery.

Discussion

The main goal of the present study was to document the existence of unilateral representational neglect in the absence of visual perceptual neglect, thereby indicating that representation and perception can be dissociated in this form of deficit (Brain, 1941; Anderson, 1993; Guariglia et al., 1993). The patient we have described appears to offer a body of evidence that is entirely consistent with this main goal. He had no apparent perceptual neglect from two months after the stroke, and as far as we were able to discern from the reports of relatives and medical and nursing staff, there were no signs of perceptual neglect from the onset of his stroke. Yet he had great difficulty in reporting details from the left side of an imaged representation based either on tactile or on visual information. This was true when he was asked to generate mental images drawn from information in his long-term memory. It was also true when he attempted to generate images from novel visual material which had been learned (long-term memory) or which had been seen only seconds before (short-term memory). Conversely, perceptual neglect has been reported in a number of patients in the absence of representational neglect (e.g. Bartolomeo et al., 1994). Therefore it is highly unlikely that patient NL would have suffered from perceptual neglect at the onset of his stroke but then have recovered from this relatively quickly while continuing to be affected by representational neglect.

The site of the patient's lesion in the right parietal lobe is in agreement with the proposal by Farah et al. (1989) and Kosslyn (1987, 1991) that the parieto-temporal region is a possible locus for the neural structures involved in mental imagery. However the patient reported by Guariglia et al. had a lesion in the frontal lobe, and therefore the neuroanatomical mappings for imagery are by no means clear (Marshall and Halligan, 1993). Also, the dissociation found between perception and imagery in both patients appears to present a challenge to theories of imagery and of visuo-spatial working memory which assume that perception and imagery rely on overlapping cognitive systems. This challenge is however more apparent than real. In neither theory do the cognitive systems completely overlap, thereby allowing scope for aspects of imagery to be impaired while perception is intact and vice versa. So too, the neural structures and pathways that have been identified as being responsible for visual perception do appear to project on to the neuroanatomical areas linked respectively with object identity and object location (Ungerleider and Mishkin, 1982) and then to a region of the posterior parietal cortex (e.g. Stein, 1992; Zeki and Shipp, 1988), which as we have mentioned above, has been linked to imagery. Therefore a lesion in the "imagery areas" might affect mental imagery but leave much of perception intact, while a more extensive lesion in the same region might affect both perception and imagery.

A second goal of this study was to explore whether the concept of visuo-spatial working memory could offer a vehicle for a theoretical account of representational neglect (with or without perceptual neglect). It is clear that the representational deficits in patient NL could not have arisen as a result of impoverished visual perceptual input, because for several of the tests the necessary information was drawn from information encoded in long-term memory before the stroke. Nor could the deficit be the result of a loss of information stored in long-term memory because he could access visual details of familiar piazzas simply by changing his imagined viewpoint. This finding suggests that all of the information was available but not all of it was directly accessible as it is in the healthy brain. Consistent with the speculation by Baddeley and Lieberman (1980) mentioned in the introduction, these observations point to a deficit in the activation of a part of the "mental screen" where the image would be represented or to a deficit of the process of scanning this "screen".

Information relevant to this argument is provided by NL's performance on the Brooks (1967) matrix task which is thought to tap the resources of visuo-spatial working memory (however see Salway and Logie, 1995, for a caveat). NL performed very poorly on the spatial version of this task, with scores close to floor. Notably he performed extremely well when he had stimulus support available in the form of the blank matrix, and did not have to rely heavily on visuo-spatial working memory. This effect of stimulus support also can account for NL's normal performance on the Corsi Blocks task in which the blocks themselves are visible throughout. Stimulus support is thought to reduce rather than to remove the temporary memory load, because NL would still have to retain the sequence of blocks. This account parallels the positive effects of stimulus support (e.g. a drawing or other physical stimulus) on working memory which have been shown in a range of imagery tasks with normal subjects (Chambers and Reisberg, 1985, 1992; Pearson, Logie and Green, 1996; for a review see Cornoldi, Logie, Brandimonte et al., 1996). Therefore visuo-spatial working memory can be supported by perception but perception is not as closely linked to working memory or to mental imagery as has widely been assumed. Hence, the enhanced performance with stimulus support found in normal subjects is consistent with the higher scores on Corsi block performance found in NL when the blocks were available to provide stimulus support. In sum, NL's deficit can be interpreted as suggesting that he has damage to important aspects of visuo-spatial working memory but that impairments are not apparent when he has support from his intact perceptual system.

The hypothesis that representational neglect reflects a deficit in visuo-spatial working memory applied not just to the impairment of our own patient NL, but also to patients who suffer from both perceptual and representational neglect. For example, the patients reported by Bisiach and Luzzatti (1978) showed symptoms of perceptual neglect on some tests. However, when they failed to describe items on the left side of the "Piazza del Duomo" as imagined from a given vantage point, their representations were derived from knowledge stored in long-term memory prior to their brain damage. Therefore their difficulty with the imaged left side of Piazza del Duomo could not have arisen from an impairment of their visual perceptual abilities at the time of the test. We are left with the inevitable conclusion that there was damage to the system responsible for generating and maintaining the temporary visual mental representation or for extracting information from that representation. That is, the patients had damage to their visuo-spatial working memory system which was independent of any damage to their perceptual system.

How might we further specify the nature of the deficit in such a system? It is worth stressing that the visuo-spatial working memory hypothesis of neglect has never been fully developed. One possible reason for this is that until recently the characterization of visuo-spatial working memory was poorly specified, although this situation is now changing (e.g. Smyth and Pendleton, 1989; Hanley et al., 1990, 1991; Logie, 1995; Salway and Logie, 1995; Quinn and McConnell, 1996). Logie (1995, Reisberg and Logie, 1993) has argued that visuo-spatial working memory may be further fractionated into a partnership of two specialised components, one referred to as the "visual cache" which stores information about visual form and colour. The second component deals primarily with spatial and movement information and is referred to as the "inner scribe". This latter partner in the system is thought to rehearse the contents of the visual cache, to transfer information from the visual cache to the central executive, and to be closely involved in the planning and execution of body and limb movements. Damage to the visual cache would permit the inner scribe to rehearse or transfer only an impoverished representation. On the other hand, damage to the inner scribe would cause great difficulty in retaining visual information, in transferring visual information to other parts of working memory such as the central executive, and in carrying out tasks that require retention of sequences of movements in the absence of stimulus support. Within this framework, NL's performance with the various versions of the Brooks' (1967) task could be interpreted as pointing to impairment of the visual cache. For example, he could retain the spatial sequence, possibly in an intact inner scribe, when he had support in the form of the blank matrix for a damaged visual cache. His difficulties with the "piazza" tests and other tests of mental representation are also consistent with the idea that information was available in the visual cache but not to the rest of working memory. This interpretation is speculative, but it does offer a coherent framework within which to understand the nature of representational neglect whether or not it is accompanied by perceptual neglect.

This interpretation requires a revision of the visuo-spatial working memory hypothesis to account for how information becomes available to working memory. The most widely held assumption is that working memory serves as a form of gateway between perceptual input and long-term memory. This assumption was explicit in the early models of short-term memory (Atkinson and Shiffrin, 1968), is maintained in most introductory textbooks on memory (e.g. Gross, 1992; Atkinson, Atkinson, Smith et al., 1993; Searlman and Hermann, 1994), is a feature of Kosslyn's theory of visual imagery (e.g. Kosslyn, 1991), and is implicit in the working memory model (e.g. Baddeley, 1986). However the "gateway" model of working memory would have difficulty interpreting representational neglect because it does not depend upon perceptual input. Moreover perceiving and interacting with the environment demands the generation of mental representations of that environment. Such representations would incorporate the location of various objects in relation to one another and the location of objects in relation to the observer. Thus when retrieving information about objects or scenes held in long-term memory, the mental representation contains information about the scenes such as when they were previously experienced, the names of objects depicted and so on (e.g. Chambers and Reisberg, 1985, 1992; Reisberg and Logie, 1993; Cornoldi et al., 1996). In this sense the contents of working memory necessarily comprise some of the products of perception and subsequent mental processing, including the semantic properties. Working memory can then be considered as a multi-component workspace which stores and manipulates information which has been activated from the long-term store, rather than as a primary route into long term memory from the environment. Therefore working memory is better viewed as a system accessed by information stored in long term memory and by sensory input only *after* that input has been processed by long term memory (for more detailed discussion of this argument see Logie, 1995, 1996; Ellis, Della Sala and Logie, 1996).

Moreover, if we see the visual cache and inner scribe as temporary visual and spatial stores which are used either after perceptual input has been removed or when visuo-spatial information is extracted from long-term memory, then this offers an account of the distinction between perception and mental representation.

Experimental support for this view comes from a wide variety of sources (e.g. Ellis, Flude and Young, 1987; Marshall and Halligan, 1988; Farah, Monheit and Wallace, 1991; Berti and Rizzolatti, 1992; McGlinchey-Berroth, Milberg, Verfaellie et al., 1993; Vallar, Rusconi and Bisiach, 1994b) which have demonstrated that on-line processing of information is accomplished by means of access to stored knowledge in long-term memory, without the information necessarily being available within working memory. For example, McGlinchey-Berroth et al. (1993) gave two neglect patients a lexical decision task, with word or non-word letter strings presented centrally. Prior to the presentation of the letter strings' pictures appeared in either the right or the left visual field. Lexical decision times for real words were faster when the pictures (presented in either hemifield) matched the meaning of the word. Therefore the pictures, even on the neglected side, appeared to activate relevant semantic information in long term memory and the level of activation was sufficient to affect on-line cognition for lexical decisions.

These data, together with those cited above, are consistent with the view that perceptual input is not processed in working memory prior to long-term memory access. These data are inconsistent with the idea of working memory as a form of gateway, as are data from the substantial literature on normal adults demonstrating priming effects (see e.g. Roediger and McDermott, 1993) and pre-conscious priming effects (e.g. Marcel, 1983a, 1983b). It is worth noting that the view that the contents of working memory comprise activated information from long-term memory already appears in alternative models of working memory (e.g. Cowan, 1988; Hasher and Zacks, 1988; Just and Carpenter, 1992; for a detailed discussion see Richardson, Engle, Hasher et al., 1996).

How successfully can this modified form of visuo-spatial working memory be applied to unilateral neglect? As stated above, it is by no means uncommon to find patients who have both perceptual neglect and representational neglect. One possible account of their deficit is that, as a consequence of their perceptual neglect, perceptual input reaches the long-term store but input from the left side is absent or activated less efficiently. The asymmetry in the levels of activation would then be passed on to the on-line representation system in working memory which (we would argue) comprises the visual cache, the inner scribe and the central executive of working memory. This could explain why representational deficits are so often associated with perceptual ones. Indeed on this view it would be possible in principle for a patient to show evidence of a representational deficit arising entirely from perceptually based neglect, that is with unilaterally impoverished perceptual input being dealt with by an intact representational system. Whether this form of dissociation is detected depends on the range of tasks given to the patient, since a representational impairment can only be revealed by tasks which present information not known to the patient before onset of the lesion.

Representational neglect cannot only be the result of a perceptual deficit because, in some patients, the representational problems arise in tasks that require the retrieval of information acquired prior to the onset of the lesion (e.g. Piazza del Duomo test). Moreover our patient NL shows representational neglect in the absence of perceptual input difficulties.
The same appears to be true of the Guariglia et al. (1993) patient.

Our hypothesis would run into difficulty were a patient to be found showing perceptual neglect in the absence of representational neglect for material presented after onset of the lesion. Some studies (Anderson, 1993; Bartolomeo et al., 1994) described patients who showed left neglect in perceptual tasks but not in imagery tasks. Anderson reported an instance of a clockface drawing in which the patient's performance was better with eyes shut (imagery task) than with eyes open (perceptual task) (see also Chedru, 1976, and Mesulam, 1985). Bartolomeo et al. reported a group of 14 out of 60 patients who performed poorly on traditional perceptual tasks (such as line cancellation) but who did not show representational neglect during descriptions from memory of familiar piazzas, countries and towns. Although these results suggest that perceptual neglect can be found in the absence of representational impairment, Anderson's (1993) and Bartolomeo et al.'s (1994) clinical cases did not show the dissociation between perceptual and representational processing that is the focus of our discussion. In fact, information used in the imagery tasks had been encoded prior to the onset of the brain lesion, and information from the left visual field would have been stored normally in long-term memory. The crucial test would be to ask the patients to describe from memory an unfamiliar pattern or array of objects which they had just seen. As far as we are aware no patient has yet been described showing perceptual neglect in the absence of representational neglect on this kind of task (Marshall and Halligan, 1993).

Marshall and Halligan (1992) argued that the understanding of neglect suffered from a lack of explanatory constructs due largely to the range of dissociations based on test performance. The working memory approach does not provide a complete account of the many behaviours that have been classified as different forms of neglect. However, the data from patient NL coupled with a reconsideration of previously reported cases of neglect suggest that visuo-spatial working memory may play an important role in and may offer the basis for an account of unilateral representational neglect.

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