

Looking while imagining

The influence of visual input on representational neglect

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Abstract—*Background:* Subjects with hemispatial neglect often exhibit representational neglect: a failure to report details from the left side of mentally visualized images. This failure could reflect impaired ability to generate the left side of the mental image, or it could reflect failure to explore the left side of a normally generated mental image. When subjects with hemispatial neglect look at pictures or drawings, their attention tends to be drawn to objects on the right side, thereby aggravating their failure to explore the left side. If representational neglect represents a failure to explore the left side of a normally generated by blindfolding, which removes the attention-catching right-sided stimuli. However, if representational neglect represents a failure to generate the left side of the mental visual image, then blindfolding should have little impact on reporting of details of the image. *Methods:* To determine which of these explanations is correct, we asked eight normal participants and eight brain-damaged patients with left representational neglect to imagine the map of France and to name as many towns as possible in 2 minutes. In different sessions, participants performed the task with eyes open or while blindfolded. *Results:* Normal participants mentioned more towns while blindfolded than with vision, thus suggesting a distracting effect of visual details on mental imagery. Patients with neglect, however, showed no appreciable effect of blindfolding on reporting of details from either side of mental images. *Conclusion:* Representational neglect may represent a failure to generate the left side of mental images.

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Representational neglect has been ascribed to a failure to generate or maintain a normal representation of the contralesional side of mental images.¹⁻³ Representational neglect is commonly assessed by requiring subjects to draw objects from memory⁴ or to name the towns or the countries on an imagined map.^{5,6} For example, when subjects with hemispatial neglect are asked to evoke mentally the map of France, they may omit to mention the towns located on the left part of the map,^{6,7} thus suggesting an amputation of the left part of their mental representation of space.^{1,8} An alternative explanation is that the mental image of contralesional space was not lacking, but rather that it was not adequately explored. This explanation is consistent with a hypothesis postulating that visual mental imagery involves some of the attentional-exploratory mechanisms that are employed in visual behavior,^{9,10} in particular, an inability to direct attention to areas of imagined space.^{1,11} The positive influence of head position,¹¹ sensory manipulations^{6,12,13} and prismatic visuomotor adaptation^{14,15} (all of which might be expected to affect exploratory behavior but not the generation of a mental image) on representational neglect in a pure imaging task fits well with this explanation.

When patients with neglect were asked to perform a drawing from memory task,^{4,16-18} with or without blindfolding, left neglect was decreased and even eliminated by blindfolding. These results suggest that visual feedback may exacerbate representational neglect and support the hypothesis that engaging attention through visual input can influence the processing of visual imagery.¹⁰ However, even in the blindfolded state, such tasks incorporate a major intentional component that underlies the act of drawing itself as well as the ongoing dynamic process involved in repeatedly comparing what is imagined to have been drawn with the original mental image template. This intentional component could serve to normalize an originally defective visual mental image. Can the presence or absence of visual input influence representational neglect in a similar way in the absence of such an intentional component? The aim of the present study was to answer this question.

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Table 1 Demographic features, clinical and CT assessed lesion site of neglect patients

Patient	Age/sex	Hemiplegia	Hemi-anesthesia	Hemianopia	Ocular and cephalic deviation	Anosognosia	Site of lesion
N1	59/M	Severe	Severe	Present	1	0	Parietal, occipital (temporal) corona radiata, insula
N2	66/M	Severe	Severe	Absent	2	1	Frontal, parietal (temporal) corona radiata, putamen
N3	47/F	Severe	Absent	Absent	1	0	Internal capsule, putamen, caudate nucleus, insula (corona radiata, frontal white matter)
N4	40/F	Severe	Severe	Present	2	0	Frontal, parietal, temporal corona radiata, insula, basal ganglia
N5	57/M	Severe	Severe	Present	1	2	Frontal, temporal, parietal, insula, basal ganglia
N6	56/M	Incomplete	Moderate	Present	1	0	Occipital, temporal (parietal)
N7	49/M	Severe	Severe	Present	1	0	Frontal (temporal, parietal), basal ganglia
N8	59/F	Severe	Moderate	Present	2	1	Parietal, occipital

Site: parentheses indicate a minimal involvement.

Methods. We studied eight right brain-damaged patients (six men, two women, mean age 55.6 \pm 10.1 years) and eight agematched healthy subjects (five men, three women; mean age 55.1 \pm 7.5 years). All subjects were right-handed and gave informed consent. All the patients had been admitted to a neurologic rehabilitation unit for treatment of left hemiplegia. Clinical features and CT scan data are described in table 1. Rightward head and eye deviation were rated on a 4-point scale: score 0 = no deviation; score 1 = intermittent deviation; score 2 = mild deviation that the subject was able to overcome with verbal instruction; score 4 = severe deviation that the subject was unable to overcome even with verbal instruction. Anosognosia for motor impairment was assessed using 4-point scale.¹⁹

All the patients showed a extensive unilateral lesion. Etiology was always vascular, ischemic in six cases and hemorrhagic in the two other cases. None of subjects had impaired arousal, confusion, dementia, or psychiatric disorders. At the time of examination, 1 month post-onset, all patients showed a marked left-sided visuo-spatial neglect defined by several tests: a line bisection task,²⁰ a line and star cancellation task,^{21,22} and reading a text and writing under dictation. All the patients also demonstrated left neglect on drawing from memory (a daisy and a clock) and on copying a daisy and a Gainotti drawing.²³ At the time of testing, only three of eight subjects (N2, N5, and N8) showed mild anosognosia.

Each subject was asked to mentally visualize the map of France as if he or she could see the map in front of him or her in his or her mind in two conditions: with eyes closed or eyes open. To help participants, they were asked to remember the map of France that they had learned during their first school period or to remember the weather forecast map featured each on television or in the newspapers. Participants had to list all the towns that they could "see" in 2 minutes.24 No instruction was given concerning the direction of mental scanning or the orientation of the mental map.²⁴ Half of the subjects began with the eyes-closed condition, whereas the remaining half proceeded in the reverse order. Responses were recorded in two ways: i) mean total scores, indicating the number of towns named, and total scores were analyzed with a two-way analysis of variance (ANOVA) (subject x condition); ii) mean left- and right-sided scores defined by the position of reported towns on the two halves of the map. Towns located inside a 75-km stripe centered on a vertical meridian line (linking Lille to Perpignan) were not taken into account (middle score). Left-right scores were analyzed with a three-way ANOVA (subject x condition x side). To have a better estimate of the location of named towns on the map in the two experimental conditions, we also measured the distance between each named town and the vertical meridian line. The distances were measured on a map of France (scale: 1/5,000,000; 1 cm = 50 km) on which all the towns that were named by the subjects were plotted. A positive value indicates a town located to the right side of the vertical meridian line and a negative value indicates a town to the left of the meridian line. Comparisons of distances were performed with two non-parametric tests: the Kruskal-Wallis ANOVA with one factor (subject or condition) and the median test, which simply counts the number of cases in neglect and healthy controls that fall above or below the common median, and computes the χ^2 value for the resulting 2×2 samples contingency table. If healthy subjects and neglect patients have identical medians, we expect approximately 50% of all cases in each sample to fall above (or below) the common median.

Results. Individual data are summarized in table 2. Healthy subjects had symmetrical scores. For all patients, the left-sided score was less than the right-sided score in both conditions, thus suggesting a deficit in image generation. To estimate more accurately the location of named towns, they were placed on a tracing of a map of France (figure 1). In healthy subjects, the reported towns are distributed over the entire map and in aggregate they create a complete map of France (figure 1A). This is consistent with the idea that performance relied on the exploration of an inner image. In patients with neglect, the named towns were placed mainly on the right half of the map, which, however, looks like the right side of the map produced by healthy subjects. This suggests a fully spared representation on the right side. However, the defective left half of the maps imagined by patients with hemispatial neglect suggests a left representational deficit (figure 1B). Notably, patients with neglect never named a town more than once, whatever its location.

In healthy subjects, mean total scores were 225 in the eyes-open condition and 259 in the eyes-closed condition, whereas in patients with neglect, the mean total scores were similar in both conditions (145 and 150). ANOVA revealed that the subject factor as well as the condition factor were significant ($F_{1,7} = 9.31$; and $F_{1,7} = 12.36$) because more towns were mentioned in eyes-closed condition (25.56 vs 23.13), and patients with neglect listed less towns than controls (18.44 vs 30.25).

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Table 2 Left-sided, right-sided, and total scores of neglect patients (N1 to N8) and mean scores of healthy subjects in two conditions of evocation of the map of France (eyes open and eyes closed)

	Eyes	s-open condit	ion		Eyes-closed condition					
Patient	Left sided	Middle	Right sided	Total	Left sided	Middle	Right sided	Total	First	
N1	1	3	7	11	2	4	9	15	Eyes open	
N2	0	1	16	17	0	2	14	16	Eyes open	
N3	2	4	12	18	1	2	22	25	Eyes closed	
N4	4	4	13	21	6	2	10	18	Eyes open	
N5	5	4	8	17	2	6	7	15	Eyes closed	
N6	2	4	21	27	1	5	21	27	Eyes closed	
N7	6	3	12	21	0	3	16	19	Eyes closed	
N8	0	2	11	13	0	2	13	15	Eyes open	
Mean	2.5	3.1	12.5	18.1	1.5	3.3	14.0	18.8		
Controls	11.1	6.1	10.9	28.1	11.4	8.6	12.4	32.4		

In healthy subjects, the mean left- and right-sided scores were 11.13 and 10.88 in eyes the eyes-open condition and 11.38 and 12.38 in the blindfolded condition, whereas in patients with neglect, these scores were statistically different (2.50 and 12.50 in the eyes-open condition and 1.50 and 14.00 in blindfolded condition). ANOVA thus revealed no significant effect of condition or side (F < 1) in healthy subjects. To check that blindfolding did not affect the evocation of towns located in the right part of the map, an additional ANOVA was performed on these items. No significant difference was found (F_{1,7} = 1.05).

In patients with neglect, three-way ANOVA revealed a

significant side effect ($F_{1,7} = 35.71$) but no condition effect. Moreover, our data suggest that the blindfolded condition slightly reduced the left-sided total score (2.5 in the eyesopen condition and 1.50 in the blindfolded condition) and slightly increased right-sided total score (12.50 in the eyesopen condition and 14.00 in the blindfolded condition) in patients with neglect. However, these changes were marginal as no significant condition x side interaction was found ($F_{1,7} = 1.67$).

In healthy subjects, Kruskal-Wallis ANOVA did not reveal a significant condition-related difference ($H_{1,407} = 0.15$), suggesting that the distribution of responses was



Figure 1. Mental evocation of map of France in eight healthy-subjects (A) and eight patients with neglect (B) with eyes open and eyes closed. Each circle indicates the location of named town on a tracing of the map (scale: 1/5,000,000; 1cm = 50 km). For each town, the size of the circle reflects the number of repetitions for all healthy subjects and patients with neglect.

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A: Healthy subjects



Figure 2. Distribution of named towns according to their position (in millimeters) relative to the vertical meridian line, measured on a map (scale: 1/5,000,000; 1 cm = 50 km) in eight healthy subjects (A) and eight patients with neglect (B) with eyes open (solid line) and eyes closed (dotted line). The vertical broken line is the position of the median.

similar in the two conditions. In addition, the median was close to the vertical meridian line and did not differ significantly in the two conditions: (median = 0 (range -1,250 to 670), median test $\chi^2 = 0.35$) (figure 2). These results suggest a symmetrical exploration of the map by normal subjects. Moreover, for any given deviation from midline, the number of towns named by normal subjects was always higher than the number named by patients with neglect except for the single sector range: 200 to 400 (figure 2). In patients with neglect, there was also no significant condition-related difference (Kruskal-Wallis ANOVA $H_{1,286} = 0.07$). The median was shifted toward the right side in both conditions (median = 279.5 [range -1250 to 650] in the eyes-open condition and median = 277.5 [range -1250 to 651] in the blindfolded condition) (figure 2B).

Finally, comparison of normal subjects with patients with neglect revealed a significant shift of the distribution and the median in patients with neglect in both the eyesopen condition (Kruskal-Wallis ANOVA H_{1,360} = 17.65; median test χ^2 = 30.65) and the blindfolded condition (Kruskal-Wallis ANOVA H_{1,407} = 35.48; median test χ^2 = 46.55).

Discussion. We wondered whether visual input might increase representational neglect as it increases visual neglect.⁴ The performances of the healthy subjects on the imagery task clearly showed a symmetrical access to the geographic knowledge when they were required to build a visual image of the map, whatever the condition, suggesting that the suppression of vision did not affect this access. However, in the blindfolded condition, the total number of named towns increased. This suggests that the lack of visual information from the environment improved the mental evocation, perhaps because blindfolded subjects were distracted by "real" visual items. During the task, the whole of the map was scanned, as suggested by the topographic distribution of the towns, consistent with a similar result found in a previous study.²⁴ The strategy of evocation appeared to rely on some kind of mental exploration, i.e., on an inner visual scanning.

The performance of patients clearly showed left representational neglect when they were asked to evoke mentally the map of France. Neglect affected the left side of the mental image, suggesting a distorted representation of the map, similar to that previously reported in a series of patients.^{6,7,15,24} In our patients, as in previous studies, the same side of space (left) was affected in mental and physical (extrapersonal) spaces. A similar co-occurrence of representational neglect with visual neglect has been reported in other group studies.^{5,7,25} Nevertheless, dissociations between representational and visuospatial neglect have been reported: visuospatial neglect in the absence of representational neglect, ^{5,18,26} representational neglect without visuospatial neglect,²⁶⁻²⁹ and even right-sided peripersonal and personal visuospatial neglect and left-sided representational neglect.30

Our patients displayed a rightward inner exploration bias. In both the eyes-open and blindfolded conditions, the retrieval and generation from long-term memory of an inner image of the map did not succeed in providing topographic information about towns on the western part of the map, but yielded normal performance on the middle and eastern parts of the map. In a task requiring only visual imagery, visual input did not influence the mental representation of space. The present findings contrast with the effects of visual feedback and visual context demonstrated in visuomotor tasks, such as drawing from memory. For example, a study reported a patient with neglect who displayed object-centered neglect with the eyes open, which disappeared with the eyes

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closed.¹⁸ In another study, a similar pattern of performance was reported in three of the six patients with neglect.⁴ In this study, five subjects with neglect showed improved drawing symmetry when blindfolded, reflecting both an increase in the extent and the number of details on the left side of the drawing and a reduction of the extent of the right. These results suggest that the attentional capture exerted by the right-sided details of drawings that subjects were producing may be reduced in the absence of visual input, thus facilitating a leftward orienting of attention. However, no similar modification of performances was observed in our patients in a pure mental imagery task during suppression of vision. The left representational neglect remained unchanged as did the number and location of named towns on the right half of the map. These findings run counter to the prediction that "the suppression of visual guidance will dramatically reduce what looks like representational neglect."⁴ It must, however, be noted that visual input was not relevant in our task, which involved pure visual imagery, whereas visual feedback regarding right-sided details involved in the drawing task was essential to performance of this task. Task-relevant visual details might be more effective in capturing patients' attention.³¹ It may also be that visual input influences performance on spatial representation tasks only when these tasks involve a manual response, i.e., an interaction between neural processes supporting visual representation and action. Even in the blindfolded state, such tasks incorporate a major intentional component that underlies the act of drawing itself as well as the ongoing dynamic process involved in repeatedly comparing what is imagined to have been drawn with the original mental image template. This intentional component could serve to normalize an originally defective visual mental image.

A recent report of a patient with pure representational neglect and poor performance on a visuospatial working memory task²⁸ suggested that the inability to build, activate, or explore the mental representation of left hemispace could result from a visuospatial working memory deficit.³² This account was explored in a recent study of 10 right-brain damaged patients with representational and very mild perceptual neglect. Patients were asked to recall immediately the names of objects presented in fourobject visual displays that had been placed directly in front of them.³³ They recalled many more rightsided than left-sided objects. This result could have been explained either by a failure of learning and generation of a visual image (working memory) of the objects in left hemispace or by failure to direct attention to left hemispace in the course of reporting what they remembered seeing. However, when the subjects were asked to recall the objects as they would appear when viewed from the opposite direction, their recall of objects in the left hemispacenow in the imagined right hemispace-did not improve, indicating impairment in their original

learning of the objects in the left hemispace; if their deficit had been in directing attention, their performance in the imagined right hemispace would have been normal. In addition, their recall of objects in the right hemispace—now the imagined left hemispace fell to the level of their original performance in the left hemispace. These results, in aggregate, are far more consistent with a working memory/image generation defect account of representational neglect than they are with a directed attention defect account. Our study provides further evidence in support of the working memory/image generation account through a task that did not require either learning of novel visual arrays or mental visualization from a different perspective. Furthermore, the fact that our subjects never repeated recalled cities in either condition suggests that they did not mentally "revisit" the same locations³⁴ and thus had a hemispace-specific deficit and not a generalized deficit in visuospatial working memory.

References

- Bisiach E, Luzzatti C. Unilateral neglect of representational space. Cortex 1978;14:129–133.
- Bisiach E, Berti A. Dyschiria. An attempt at its systemic explanation. In: Neurophysiological and neuropsychological aspects of spatial neglect, Jeannerod M, ed. Amsterdam: North Holland, 1987:183–201.
- Berti A. Cognition in dyschiria: Edoardo Bisiach's theory on misconception of space and consciousness. Cortex 2004;24:275–280.
- Chokron S, Colliot P, Bartolomeo P. The role of vision on spatial representations. Cortex 2004;40:281–290.
- Bartolomeo P, D'Erme P, Gainotti G. The relationship between visuospatial and representational neglect. Neurology 1994;44:1710–1714.
- Rode G, Perenin MT. Temporary remission of representational hemineglect through vestibular stimulation. Neuroreport 1994;5:869–872.
- Rode G, Perenin MT, Boisson D. Négligence de l'espace représenté: mise en évidence par l'évocation mentale de la carte de France. Rev Neurol (Paris) 1995;151:161-164.
- 8. Bisiach E, Capitani E, Luzzatti C, Perani D. Brain and conscious representation of outside reality. Neuropsychologia 1981;19:543–551.
- Thomas NJT. Are theories of imagery theories of imagination? An active perception approach to conscious mental content. Cogn Sci 1999;23: 207-245.
- Bartolomeo P, Chokron S. Can we change our vantage point to explore imaginal neglect? (Commentary on Pylyshyn: Mental imagery: in search of a theory). Behav Brain Sci 2002;25:184–185.
- Meador KJ, Loring DW, Bowers D, Heilman KM. Remote memory and neglect syndrome. Neurology 1987;37:522–526.
- Geminiani G, Bottini G. Mental representation and temporary recovery from unilateral neglect after vestibular stimulation. J Neurol Neurosurg Psychiatry 1992;55:332–333.
- Vallar G. Modulation of the neglect syndrome by sensory stimulation. In: Parietal lobe contribution to orientation in 3D space, Their P, Karnath HO, eds. Heidelberg: Springer-Verlag, 1997:555-579.
- Rode G, Rossetti Y, Li L, Boisson D. Improvement of mental imagery after prisms exposure in neglect: a case study. Behav Neurol 1999;11: 251-258.
- Rode G, Rossetti Y, Boisson D. Prism adaptation improves representational neglect. Neuropsychologia 2001;39:1250–1254.
- Chedru F. Space representation in unilateral neglect, J Neurol Neurosurg Psychiatry 1976;39:1057–61.
- Mesulam MM. Principles of behavioral neurology. Philadelphia: FA Davis, 1985.
- Anderson B. Spared awareness for the left side of internal visual images in patients with leftsided extrapersonal neglect. Neurology 1993; 43:213-216.
- Bisiach E, Vallar G, Perani D, Papagno C, Berti A. Unawareness of disease following lesions of the right hemisphere: anosognosia for hemiplegia and anosognosia for hemianopia. Neuropsychologia 1986;24:471– 482.
- Schenkenberg T, Bradford DC, Ajax ET. Line bisection with neurological impairment. Neurology 1980;30:509–517.
- Albert MLA. A simple test of visual neglect. Neurology 1973;23:658– 673.
- Wilson BA, Cockburn J, Halligan PW. Behavioral Inattention Test. England: Thames Valley Test Company, 1987.

436 NEUROLOGY 68 February 6, 2007

- Gainotti G, Messerli P, Tissot R. Qualitative analysis of unilateral spatial neglect in relation to the laterality of cerebral lesions. J Neurol Neurosurg Psychiatry 1972;35:545-550.
 Rode G, Rossetti Y, Perenin MT, Boisson D. Geographic information
- Rode G, Rossetti Y, Perenin MT, Boisson D. Geographic information has to be spatialised to be neglected: a representational neglect case. Cortex 2004;40:391–397.
- Bisiach E, Luzzatti C, Perani D. Unilateral neglect, representational schema and consciousness. Brain 1979;102:609–618.
- Coslett HB. Neglect in vision and visual imagery: a double dissociation. Brain 1997;120:1163–1171.
- Guariglia C, Padovani A, Pantano P, Pizzamiglio L. Unilateral neglect restricted to visual imagery. Nature 1993;364:235–237.
- Beschin N, Cocchini G, Della Sala S, Logie R. What the eyes perceive, the brain ignores: a case of pure unilateral representational neglect. Cortex 1997;33:3-26.

- 29. Ortigue S, Viaud-Delmon I, Annoni JM, et al. Pure representational neglect after right thalamic lesion. Ann Neurol 2001;50:401–404.
- Beschin N, Basso A, Della Sala S. Perceiving left and imagining right: dissociation in neglect. Cortex 2000;36:401–414.
- Ptak R, Schnider A. Reflexive orienting in spatial neglect is biased towards behaviourally salient stimuli. Cereb Cortex 2006 16:337-345.
- Baddeley AD, Lieberman K. Spatial working memory. In: Attention and performance VIII, Nikerson RS, ed. Hillsdale, NJ: Lawrence Erlbaum Associates, 1980:521–539.
- Della Sala S, Logie RH, Beschin N, Denis M. Preserved visuo-spatial transformations in representational neglect. Neuropsychologia 2004;42: 1358–1364.
- Husain M, Mannan S, Hodgson T, Wojciulik E, Driver J, Kennard C. Impaired spatial working memory across saccades contributes to abnormal search in parietal neglect. Brain 2001;124:941–952.

Neuro Images



Figure. MRI scans. An area of abnormal signal, probably of demyelinating origin, is evident in the upper medulla and pons. No other signal abnormalities are evident within the CNS; in particular, the cervical cord is spared (B).

VIDE0Pathologic startle followingbrainstem lesion

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The startle reflex is a motor response that originates in the lower brainstem.¹ Abnormal symptomatic startle can be secondary to lesions in the startle pathway, involving brainstem and

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spinal cord.² A 56-year-old woman developed an acute demyelinating lesion of unknown origin in medulla oblongata (figure, A–D), causing dizziness and bilateral sensory impairment with paresthesias. No tongue weakness, myoclonus, or symptoms of restless leg syndrome were present. When the symptoms remitted, she developed a severe symptomatic startle response. Pathologic startle was elicited by sensory—especially acoustic—stimuli (video, see the *Neurology* Web site at www. neurology.org). Startle was bilateral and the EMG burst duration, recorded with surface deltoid EMG, ranged from 500 to 1,200 msec. Startle was not responsive to pharmacologic treatment (benzodiazepines and carbamazepine) and was disabling for the patient.

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 Cruccu G, Deuschl G. The clinical use of brainstem reflexes and handmuscle reflexes. Clin Neurophysiol 2000;111:371–387.

 Jankelowitz SK, Colebatch JG. The acoustic startle reflex in ischemic stroke. Neurology 2004;62:114–116.