**REVIEW ARTICLE**

Topographical disorientation: a synthesis and taxonomy

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**Summary**

Over the last century, several dozen case reports have presented ‘topographically disoriented’ patients who, in some cases, appear to have selectively lost their ability to find their way within large-scale, locomotor environments. A review is offered here that has as its aim the creation of a taxonomy that accurately reflects the behavioural impairments and neuroanatomical findings of this literature. This effort is guided by an appreciation of the models of normative way-finding offered by environmental psychology and recent neuroscience research. It is proposed that several varieties of topographical disorientation exist, resulting from damage to distinct neuroanatomical areas. The particular pattern of impairments that patients evidence is argued to be consonant with the known functions of these cortical regions and with recent neuroimaging results. The conflicting claims of previous reviews of this area are also considered and addressed.

**Keywords:** topographical disorientation; topographical agnosia; topographical amnesia; spatial disorientation; way-finding

**Introduction**

Some of the most compelling evidence for functional, anatomical divisions of mental operations is provided by patients with specific cognitive impairments following localized brain lesions. Over the last century, several dozen case reports have presented patients who seem to have selectively lost the ability to find their way within their locomotor environment. Of the many labels that have been applied to this impairment, *topographical disorientation* has been the most consistently adopted. If selective cognitive impairments can imply specialized neural systems, then are we to interpret cases of topographical disorientation as evidence of neural substrates specialized for the representation of large-scale, environmental space? If so, what do these disorders tell us of the organization and operation of normative way-finding abilities? The review that follows addresses these questions.

It should be made clear from the outset that way-finding is a complex, multi-component behaviour. Many different solutions (some discussed below) to the challenge of travelling from ‘A’ to ‘B’ have been offered, and there is little doubt that people, being canny problem solvers, make use of a variety of strategies in a variety of settings. Thus, the term ‘topographical disorientation’ is itself rather vague, as it is possible that disorientation could follow the loss of any one of many cognitive abilities. Furthermore, general impairments (e.g. blindness, global amnesia, dementia) can also be expected to impact way-finding. The first order of business then will be a brief consideration of the cognitive processes involved in way-finding. This effort will provide a lexicon for the impairments that topographically disoriented patients display and will be followed by a consideration of how clinical tests of disoriented patients are to be interpreted.

After a historical overview and a consideration of past frameworks, the homogeneity of topographical disorientation is considered. Is it the case that all patients who are described as ‘topographically disoriented’ possess the same impairment? If not, how do these cases differ from one another? Can patients present with selective impairments in way-finding ability? In answering these questions, the outlines of a taxonomy of topographical disorientation are formed. While this organization is guided primarily by consideration of the behavioural deficits evidenced by different patients,
the lesion sites that have been reported also play a role in the framework.

As reviews of the topographical disorientation literature have been provided before (De Renzi, 1982; Riddoch and Humphreys, 1989), including a recent, comprehensive review (Farrell, 1996), the reader may question the need for an additional treatment. There are several motivations for the current effort. Numerous additional cases have been reported since the earlier reviews. These recent reports present high-resolution radiological studies (such as CT and MRI), thus greatly improving the identification of the lesion site responsible for these deficits. More importantly, however, the organization and interpretation of the cases offered here differs in several respects from that of previous authors. Throughout this review, the proposals of previous authors will be considered and points of disagreement highlighted.

Finally, it should be noted that case review, no matter how insightful, is not hypothesis testing. The goal here is parsimony: the distillation of a century’s worth of idiosyncratic case reports into minimal taxonomy. This effort has at its disposal almost unlimited degrees of freedom. As a result, while the current review may be judged by its concordance with prevailing notions in psychology and neurobiology, the final arbiter of its accuracy must be further tests of patients and neurologically intact controls based upon the predictions of the model.

Normative way-finding

Environmental psychology is concerned with the knowledge that people acquire about the space beyond the range of their immediate perception. This field has early and often emphasized the heterogeneity of the behaviours that humans are able to perform within their extended environment. Subjects range widely in their ability to reproduce maps of their neighbourhood, make estimates of distance and direction to landmarks or find their way from one location to another. These variations have been, at times, attributed to subject variables (e.g. gender, age, length of residence), differences in environmental characteristics (e.g. density of landmarks, regularity of street arrangements) and differences in forms of knowledge acquisition (e.g. navigation versus map learning). One basic tenet of many environmental psychology studies is that these differences are to a great extent the result of differences in representation—a subject not only improves his or her knowledge of the environment with increasing familiarity, for example, but comes to represent that knowledge in qualitatively different ways with experience (Piaget et al., 1960; Appleyard, 1969; Siegel and White, 1975; Siegel et al., 1978). This shift in representation in turn supports the ability to produce more accurate, flexible and abstract spatial judgments. Specifically, a distinction has frequently been drawn between representations of the environment that are route based and those that are more ‘map-like’. This gross division has appeared under many labels [i.e. route versus configural (Siegel and White, 1975); taxon versus locale (O’Keefe and Nadel, 1978); network versus vector map (Byrne, 1982); procedural versus survey (Thorndyke and Hayes-Roth, 1982)], but generally possesses the same basic structure.

Most environmental representation is predicated upon the ability to recognize specific locations where navigational decisions are executed. This perceptual ability is termed landmark (or place) recognition and is thought to be the first ‘topographic’ ability acquired in developing infants (Piaget et al., 1960). Subjects improve in their ability to successfully identify environmental features with developmental age and have considerable between-subject agreement as to what constitutes a useful landmark (Allen et al., 1979).

Route knowledge describes the information that encodes a sequential record of steps that lead from a starting point, through landmarks, and finally to a destination. This representation is essentially linear, in that each landmark is coupled to a given instruction (i.e. go right at the old church), that leads to another landmark and another instruction, repeated until the goal is reached. Indeed, the learning of landmark-instruction paths has been likened to the learning of stimulus–response pairs (Thorndyke, 1981). While more information can be stored along with a learned route—for example, distances, the angles of turns and features along the route (Thorndyke and Hayes-Roth, 1982)—there is evidence that subjects often encode only the minimal necessary representation (Byrne, 1982).

Descriptions of route learning also emphasize its grounding in an egocentric co-ordinate frame. It is assumed that a set of transformations take place by which the retinal position of an image is combined with information regarding the position of the eyes in the orbits and the position of the head upon the neck in order to represent the location of an object with reference to the body. This is termed egocentric (or body-centred) space and is the domain of spatial concepts such as left and right. Orientation is maintained within a learned route by representing egocentric position with respect to landmark (i.e. pass to the left of the grocery store, then turn right). A final, and crucial, aspect of route-knowledge is its presumed inflexibility. Because a route encodes only a series of linear instructions the representation is fragile, in that changes in crucial landmarks or detours render the learned path useless.

Whereas route-learning is conducted within egocentric space, map-like representations are located within the domain of exocentric space, in which spatial relations between objects within the environment, including the observer, are emphasized (Taylor and Tversky, 1992). A developmental dissociation between egocentric and exocentric spatial representation has been demonstrated in a series of experiments by Acredolo indicating that these two co-ordinate frames are represented by adult subjects (Acredolo, 1977). In order to generate a representation of exocentric space, egocentric spatial decisions must be combined with an integrated measure of one’s motion in the environment. While a tree may be to my right now, if I walk forward 10 paces
and turn around, the tree will now be to my left. Though the egocentric position of the landmark has changed I am aware that the tree has not moved; the exocentric position has remained invariant. A representation of this invariance is available by combining the egocentric spatial judgements with a measure of the vector motion that was undertaken.

Because of this representational co-ordinate frame, map-like representations are thought to preserve the Euclidean relationships between places in the environment. Thus, angle and distance relations between landmarks are readily available. In addition, maps are inherently flexible. This is a feature emphasized by O’Keefe and Nadel: ‘Whereas a route specifies a starting point, a goal, and a particular direction of movement from the former to the latter, a map specifies none of these, either in its construction or its usage. It can be used with equal facility to get from any particular place to any other. Additional flexibility derives from the freedom from specific objects and behaviours. If one path is blocked another can be easily found and followed.’ (O’Keefe and Nadel, 1978, p. 87).

The observation that humans use different navigational strategies at different times has led to the investigation of the conditions under which these strategies are differentially invoked and their subsequent impact upon performance. Studies have demonstrated that, given text descriptions of a place, subjects tend to form map-like representations when the description is rooted in survey relations. When provided with a more route-based description, subjects not surprisingly form a route-based representation (Ferguson and Hegarty, 1994). Similarly, subjects provided with an aerial view map more easily generate a representation with map-like form (Thorndyke and Hayes-Roth, 1982). Features of the environment also impact representational form. Heft has suggested that in relatively undifferentiated environments (i.e. those with few landmarks), subjects tend to employ map-like representations (Heft, 1979). In environments with rich landmark detail subjects employ route-learning and use this technique preferentially (Acredolo and Evans, 1980). Yet another source of variance in representation is developmental age and environmental familiarity. Piaget and colleagues proposed that children develop different cognitive representations of space as they age, progressing from initial landmark recognition to egocentric route learning and finally to metric representations of space in the exocentric frame (Piaget et al., 1960). This developmental progression has been confirmed by studies of environmental learning in children (Acredolo, 1977; Allen et al., 1979; Bremner, 1978; Cousins et al., 1983; Golledge et al., 1985; Herman et al., 1987; Garino and McKenzie, 1988). Based upon this developmental progression in children and from observations of adult performance, Siegel and White proposed that the development of environmental knowledge is characterized by qualitative shifts in the representation of space from memory for traversed routes to a more abstract, map-like representation of object location (Siegel and White, 1975). While the general prediction of this model, i.e. improvement in estimates of survey measures with increased environmental familiarity, has been confirmed in a number of studies (e.g. Thorndyke and Hayes-Roth, 1982; Cousins et al., 1983), it has also been demonstrated that subjects maintain rather rudimentary route-knowledge of their environments even after years of familiarity (Byrne, 1982).

**Interpreting the clinical tests of topographical knowledge**

An important lesson from this cursory review of environmental psychology concerns the heterogeneity of the techniques that subjects bring to bear upon navigational problems. The particular type of representation that a subject generates of his environment has been shown to be dependent upon (i) the subject’s developmental age, (ii) the duration of a subject’s experience with a particular environment, (iii) the manner in which the subject was introduced to the environment (i.e. self-guided exploration, map reading), (iv) the level of differentiation (detail) of the environment, and (v) the tasks that the subject is called upon to perform within the space. The multiplicity and redundancy of strategies that may be brought to bear upon way-finding challenges makes the interpretation of standard clinical tests of topographical orientation problematic. For example, asking a patient to describe a route in her town is not guaranteed to evoke the same cognitive processes for different routes, let alone different subjects. As these commonly employed tests of topographical orientation (i.e. describe a route, draw a map) are poorly defined with regard to the cognitive processes they require, it is always possible to provide a post hoc explanation for any particular deficit observed.

This inferential complication is further confounded by the ability of patients to store a particular representation in any one of several forms. Consider, for example, the frequently employed bedside test of sketch-map production. Patients are asked to draw a simple map of a place (e.g. their home, their town, the hospital) with the intention of revealing intact or impaired exocentric (i.e. map-like) representations of space. It is possible, however, to produce a sketch-map of a place without possessing an exocentric representation (Pick, 1993). For example, complete route knowledge of a place, combined with some notion of the relative path lengths composing the route segments, is sufficient to allow the construction of an accurate sketch-map. Thus, while a subject may be able to produce a sketch-map of a place, this does not necessarily indicate that the subject ever possessed or considered an exocentric representation of that place prior to the administration of the test (Byrne, 1982). Alternatively, it is possible that considerable experience with map representations of a place would lead a subject to develop a ‘picture-like’ representation. If, for example, a subject has had the opportunity to consult or draw maps of his home or home-town several times previously, then he might be able to draw a map of that place in the same manner that he might draw a picture of an object.
In a similar manner, impairments in one area of topographical representation might lead to poor performance in tests that ostensibly probe a different area of competence. For example, asking a patient to describe a route through a well-known place is frequently presumed to rely only upon intact egocentric spatial knowledge. However, it is entirely possible that, if producing a verbal description of a route is not a well-practiced behaviour, subjects engage in an imaginal walk along the route to produce the description (Farrell, 1996). In this case, deficits in the ability to represent and manipulate information regarding the appearance of landmarks would also impair performance. Thus, given that subjects might only generate map-like representations at the time of testing, and given that this process can be dependent upon route representations which themselves may require intact representations of environmental landmarks, it is conceivable that tertiary impairments in sketch-map production might be produced by primary impairments in landmark recognition!

How then are we to proceed in interpreting the extant topographical disorientation literature? The only possible means of gaining an inferential handle upon these disorders is to obtain additional information regarding the nature of the impairment. One simple approach is to attach credence to the patient’s description of their disability. As will be examined below, some categories of topographical disorientation give rise to rather consistent primary complaints across patients. When these reports are sufficiently clear and consonant they provide a reasonable basis for theorizing. Naturally, there are limitations to this approach as well. Patient reports might simply be wrong (Farrell, 1996); the case of De Renzi and Faglioni offers an example in which the patient’s claim of intact recognition for buildings and environmental features was at odds with his actual performance (De Renzi and Faglioni, 1962).

Additional clinical tests, with more transparent interpretations, may also be used to help interpret topographical impairments. Particularly helpful have been demonstrations of stimulus-specific deficits in visual memory and of impairments of egocentric spatial representation. While more complex clinical tests have been employed, these frequently are as subject to various interpretation as is the original patient deficit. For example, the stylus-maze task (Milner, 1965), in which the subject must learn an invisible path through an array of identical bolt-heads, has been widely applied. Despite the vague similarity of maze learning and real-world navigation, it is conceivable that failure to complete the task successfully might be due to a number of cognitive impairments that are unrelated to way-finding; indeed, neuropsychological studies that have employed this test have noted that many patients who are impaired on the stylus-maze task have no real-world orientation difficulties whatsoever (Newcombe and Russell, 1969), and vice versa (Habib and Sirigu, 1987).

The ability of patients to compensate for their deficits, and the techniques that they use, is also informative. For example, some patients have been reported to navigate by reference to an extensive body of minute environmental features, such as distinctive door-knobs, mailboxes and park benches (Meyer, 1900). As will be discussed below, this compensatory strategy speaks both to the nature of the impairment and to the intact cognitive abilities of the patient. Finally, the traditional ‘sketch-map production’ and ‘route-description’ tests can provide useful information in some situations. Consider the case of a patient who is able to generate accurate sketch-maps of places that were unfamiliar to him prior to sustaining his lesion and that he has only experienced through direct exploratory contact. In this situation, the patient must have an intact ability to represent spatial relationships (either egocentric or exocentric) to have been able to generate this representation. In a similar vein, the demonstration of intact representational skills using these ‘anecdotal’ clinical measures may be interpreted with slightly more confidence than impairments.

Initial cases and previous frameworks

Although there were earlier case reports (Foerster, 1890; Jackson, 1932; well summarized by De Renzi, 1982), a description by Meyer (Meyer, 1900) of three patients with topographical disorientation represents the first comprehensive study of the disorder. Meyer’s first patient, a 49-year-old man, presented with a left homonymous hemianopsia and severe disorientation following a vascular lesion. Despite generally intact intellect, visual perception and memory, this patient was unable to find his way in his home town or learn his way around the hospital. He was unable to describe or draw the route between his home and any of the principle public places in his town. In addition, he had great difficulty in recognizing places by their appearance, and could deduce their identity only by taking note of small details. Thus, he was only able to determine which ward within the hospital was his by looking for the black beard of his room-mate. In contrast to Meyer’s patient, the topographically disoriented patients described by Holmes and Horrax (Holmes and Horrax, 1919) and Brain (Brain, 1941) suffered from notable impairments of immediate spatial perception, such that they were unable to judge the distance and direction of objects.

Considering these cases, and their own patient (described in detail below), Paterson and Zangwill proposed that the disorder is one of ‘specific topographical agnosia’ (Paterson and Zangwill, 1945). By this they presumably meant that while patients are able to recognize broad categories of environmental information, such as spatial distances and building categories (church, school, etc.), they are unable to visually identify specific instances of these topographical features. Thus, to paraphrase Milner and Teuber, these patients were proposed to possess normal environmental percepts, stripped of their topographic meaning (Milner and Teuber, 1968).

Many authors have subsequently adopted and extended
this terminology, often arguing that instead of a topographical agnosia, the deficits are better described as an amnesia. The application of these somewhat vague terms has been, however, rather idiosyncratic. As Farrell astutely noted, Paterson and Zangwill did not provide a clear delineation of these categories in their original paper (Farrell, 1996). Subsequently, the terms have come to be applied to a variety of disorders, with different authors arguing for the primacy of agnosia or amnesia deficits in their particular group of patients without any particularly clear statement of what theoretical issues are at stake. For example, some authors interpret topographical amnesia as a deficit primarily in spatial memory and topographical agnosia as an impairment of environmental perception (Landis et al., 1986). Others have considered topographical agnosia a ‘visual–spatial’ deficit and topographical amnesia a ‘visual memory’ deficit (Cammalleri et al., 1996). Additional constructions have further subdivided the agnosias and amnesias each into separate spatial and perceptual categories (Bottini et al., 1990; Takahashi et al., 1997), while other authors have equated agnosia and amnesia impairments with losses of recognition and familiarity, respectively, for environmental features (Habib and Sirigu, 1987). Finally, some authors have recognized all of these varieties (i.e. agnosia for spatial information, agnosia for landmark information, amnesia for spatial information, etc.) (Incisa della Rocchetta et al., 1996). It is, in fact, rather difficult to provide a detailed account of the use and evolution of these terms over the last few decades, as the contexts in which they have been deployed and the models of neural organization which they tacitly embrace have seldom been carefully addressed (for gallant attempts of neural organization which they tacitly embrace models of neural organization which they tacitly embrace).

In 1985, Levine and colleagues, following in the footsteps of previous authors (Whiteley and Warrington, 1978), proposed that topographically disoriented patients may be more profitably divided into two groups: those with impairments in representing spatial visual information, and those impaired in representing object visual information (Levine et al., 1985). The authors based this proposal upon the now widely accepted model of ‘two streams’ of visual processing (Ungerleider and Haxby, 1994), which proposes that separable dorsal and ventral posterior cortical areas subserve the analysis of spatial position (‘where’) and object identity (‘what’), respectively. They also presented two patient cases, the second of which was relevant for his assertion. Case 2 was a gentleman who was rendered topographically disoriented, among other deficits, by a right parieto-occipital haematoma: ‘He could not find his way about. At 4 months after the hemorrhages, he frequently got lost in his own house. . . . Spatial imagery was severely impaired. He could not say how to get from his house to the corner grocery store, a trip he had made several times a week for more than 5 years. In contrast, he could describe the store and its proprietor.’

The patient’s deficits suggested to the authors that severe spatial disorientation could exist with intact visual (landmark) imagery and perception. They further contended based upon a review of published cases, that prosopagnosia, pathognomonic for Levine and colleagues of visual–object impairment, could be present without accompanying spatial disorientation. They then argued that different fundamental disorders might lead to impaired route following, noting that ‘prosopagnostic patients lose their way because they cannot recognize landmarks . . . [while] patients with visual disorientation recognize landmarks but cannot find their way because they do not know how to orient the body with respect to these landmarks.’ In short, Levine and colleagues suggested that topographical disorientation could result from the lesioning of two separate systems: one required for the identification of salient landmarks and one required for the representation of spatial position.

The formulation of Levine and colleagues, while offering a promising theoretical framework in which to consider the disorder of topographical disorientation, was limited as initially presented. First, while they did review the literature of prosopagnosia for anecdotal reports of visual–object and visual–spatial impairments, specific cases with demonstrated impairments in landmark recognition and demonstrably preserved visual–spatial abilities were not presented. Thus, while the review did suggest that, for example, colour vision and spatial orientation can be separately impaired, no evidence was presented for the independence of specifically topographical visual–object abilities (i.e. landmark recognition) and spatial orientation. Secondly, the authors suggested that topographical disorientation of the visual–object variety was the result of the same impairment that produces prosopagnosia, at times using the labels interchangeably. This assertion runs counter to the evidence that these two disorders can occur independently (discussed below), including, interestingly, case 1 presented by Levine and colleagues. Finally, the ‘what/where’ divisional model did not make reference to the kinds of spatial representations putatively stored in the dorsal cortex or to the selectivity of the deficits for topographical knowledge.

An alternate model presented in 1995 by Milner and Goodale (Milner and Goodale, 1995) questioned the macroscopic functional organization of the cortex that underpins the account of Levine and colleagues. In their comprehensive critique of the ‘dorsal/ventral—what/where’ model, the authors argued that both dorsal and ventral areas of posterior cortex process location and identity information, but for different purposes: the dorsal stream subserves the production of actions while the ventral stream is necessary for identification. Within the scope of topographical representation, dorsal regions maintain representations of egocentric spatial information while ventral regions subserve both landmark identity and ‘allocentric’ (i.e. exocentric)
spatial representations. In the case of the ventral system, however, it is not clear from the account by Milner and Goodale if (i) the two functions are somehow necessarily behaviourally linked, or (ii) identical neural substrates subserve both landmark and exocentric spatial functions.

Regardless, Milner and Goodale describe only a few cases of topographical disorientation, and several of these are not particularly relevant to the proposed model. A later effort by Farrell (Farrell, 1996), however, presents a comprehensive review of the topographical disorientation literature with the specific aim of comparing the model advanced by Levine with that of Milner and Goodale. Farrell argues that there is not strong evidence for the Levine model and instead finds the alternate framework of Milner and Goodale in better accord with the topographical disorientation literature. Notably, Farrell interprets the Milner and Goodale ‘perception/action’ account as proposing that the same neural substrates subserve landmark recognition and exocentric spatial representation: ‘Such a model [the perception/action model] would not predict a double dissociation between the identification of objects and knowledge of spatial relationships as they are both served by the same subsystem.’ (Farrell, 1996).

Farrell’s interpretation thus makes several predictions that distinguish it from the model of Levine and colleagues: (i) any patients with intact landmark recognition, but impaired spatial representation, should have spatial impairments confined to the egocentric sphere; (ii) patients with impaired landmark recognition but intact spatial representation should not be observed; and (iii) there should exist patients with isolated ventral lesions who are impaired on both tests of landmark identification and allocentric representation. Ultimately, Farrell’s notion that dorsal, parietal lesions produce only egocentric spatial disorientation is well supported by the literature. Below, we describe several cases of this form of ‘egocentric disorientation.’ The other predictions of the model proposed by Farrell are not, however, strongly affirmed.

First, there is little evidence for the existence of patients impaired in both landmark recognition and exocentric spatial representation following an isolated ventral lesion. Many of the cases Farrell cites as having mixed recognition and spatial deficits are unable to contradict either the model by Milner and Goodale or Levine and colleagues, as the lesion site is either unknown (De Renzi and Faglioni, 1962) or large and distributed (Paterson and Zangwill, 1945; Whitty and Newcombe, 1973; Clarke et al., 1993). Two of the cases cited within Farrell’s section on spatial disorientation in fact suffer only from impaired landmark recognition (discussed below) (Pallis, 1955; Hécaen et al., 1980), while one case cited (patient 4 of Aimard et al., 1981) presented only with impaired spatial relational knowledge following a ventral lesion. (This last case is in fact problematic for the accounts of both Farrell, and Levine and colleagues.) Farrell also correctly notes that in a few cases where isolated spatial deficits have been claimed, the patients have not been tested to verify their statements of intact visual recognition.

Secondly, and more damaging to the Milner and Goodale model, is the evidence for impaired landmark recognition in the setting of intact spatial representation. The existence of these cases is very hard to reconcile with the argument that a single neuroanatomical substrate suberves the representation of both the identity of environmental information (landmarks) and their relative positions.

A taxonomy of topographical disorientation

What, then, can we say about the organization of neural systems for way finding and the disorder of topographical disorientation? Our examination of the literature suggests a way to divide the cases that has its foundations both in the accounts of Levine and colleagues, and Farrell, but modifies and extends these frameworks. Three categories of disorientation are defined with a fourth considered as a possibility. These divisions reflect the natural ‘fault lines’ of the cases as we interpret them, and are informed by recent observations of the functional organization of the most common neuroanatomical lesion sites. The first of these categories agrees entirely with a proposal made by Milner and Goodale: lesions of the posterior parietal cortex produce a variant of disorientation that is rooted in the egocentric spatial sphere. Secondly, there is a small but intriguing set of cases that suggest that representation of ‘heading’ can be selectively impaired. These cases of ‘heading disorientation’ are particularly interesting given that the reported lesion site has been shown to contain ‘head direction’ cells in the rodent. Thirdly, we suggest that isolated lesions of the ventral cortex can produce a ‘landmark agnosia’ in which patients are unable to recognize salient environmental landmarks. While this third category is similar to that proposed by Levine and colleagues, there are important differences. In particular, we suggest that this disorder is the result of damage to a system that is actually specialized for landmark representation, as opposed to a general object recognition system. As these cases are among the most numerous and best studied, we will consider this category of topographical disorientation at greatest length. Next, some cases suggest that parahippocampal lesions can produce primarily an antero-grade deficit in topographical orientation. The difficulty, however, in distinguishing these cases from cases of landmark agnosia is discussed. We will also discuss the role of the hippocampus in topographical orientation in the human as this structure has been intensely studied with regard to navigation in the rodent. As we proceed, recent observations from functional neuroimaging studies will be considered as they relate to the categories described (Table 1).

Egocentric disorientation

The first group of patients to be considered have traditionally been labelled as ‘topographically disoriented,’ but it will be
argued that their impairments are not strictly confined to the topospheric sphere. Several cases of egocentric disorientation have been reported, and include patients MNN (Kase et al., 1977), Mr Smith (Hanley and Davies, 1995), GW (Stark et al., 1996) and the cases of Holmes and Horax (Holmes and Horax, 1919), and are well represented by case 2 of Levine and colleagues (Levine et al., 1985):

The most striking abnormalities were visual and spatial. He could not reach accurately for visual objects, even those he had identified, whether they were presented in central or peripheral visual fields. When shown two objects, he made frequent errors in stating which was nearer or farther, above or below, or to the right or left. He could not find his way about. At 4 months after the hemorrhages, he frequently got lost in his own house and never went out without a companion. Spatial imagery was severely impaired. He could not say how to get from his house to the corner grocery store, a trip he had made several times a week for more than 5 years. In contrast, he could describe the store and its proprietor. His descriptions of the route were frequently bizarre: ‘I live a block away. I walk direct to the front door.’ When asked which direction he would turn on walking out of his front door, he said, ‘It’s on the right or left, either way.’ When, seated in his room, he was blindfolded and asked to point to various objects named by the examiner, he responded [very poorly].

These patients, as a group, have severe deficits in representing the relative location of objects with respect to the self. While they are able to gesture towards visualized objects, for example, this ability is completely lost when their eyes are closed. Performance is impaired on a wide range of visual–spatial tasks, including mental rotation and spatial span tasks. It thus seems appropriate to locate the disorder within the egocentric spatial frame. Indeed, Stark and colleagues (Stark et al., 1996) have suggested that one of these patients (GW) has sustained damage to a spatial map that represents information within an egocentric coordinate system. Interestingly, these cases suggest that neural systems capable of providing immediate egocentric position information can operate independently of systems that store this information (Stark et al., 1996).

These patients are uniformly impaired in way-finding tasks in both previously familiar and novel environments. Most remain confined to the hospital or home, willing to venture out only with a companion (Kase et al., 1977; Levine et al., 1985). Route descriptions are impoverished and inaccurate (Levine et al., 1985; Stark et al., 1996) and sketch-map production is disordered (Hanley and Davies, 1995). In contrast to these impairments, visual–object recognition has been informally noted to be intact. Patient MNN (Kase et al., 1977) was able to ‘name them [objects] correctly without hesitation, showing an absence of agnosic features in the visual sphere’. Patient GW (Stark et al., 1996) ‘experienced no difficulty in recognizing people or objects’ and case 2 of Levine and colleagues (Levine et al., 1985) was able to ‘identify common objects, pictures of objects or animals, familiar faces or photographs of the faces of family members and celebrities’.

Unfortunately, these patients have not been specifically tested on visual recognition tasks employing ‘landmark’ stimuli. As noted above, Levine and colleagues report that their case 2 was able to describe a grocery store and its proprietor, but this does not constitute a rigorous test. It is possible that, despite demonstrating intact object and face recognition abilities, patients with egocentric disorientation will be impaired on recognition tasks that employ topographically relevant stimuli. Thus, until these tests are conducted, we can offer only the possibility that these patients are selectively impaired within the spatial sphere.

It seems plausible that the way-finding deficits that these patients display are a result of their profound disorientation in egocentric space. As was discussed in the introduction, route-based representations of large-scale space are formed within the egocentric spatial domain. This property of spatial representation was well illustrated by Bisiach and colleagues with a study of route descriptions in a patient with unilateral neglect (Bisiach et al., 1993). Regardless of the direction that the subject was instructed to imagine travelling, turns on the left-hand side tended to be ignored. Thus, the egocentric disorientation that these patients display seems sufficient to account for their topographical disorders. In this sense, it is perhaps inappropriate to refer to these patients as selectively topographically disoriented—their disability includes forms

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**Table 1** A four-part taxonomy of topospheric disorientation

<table>
<thead>
<tr>
<th>Lesion site</th>
<th>Disorder label</th>
<th>Proposed impairment</th>
<th>Model case</th>
</tr>
</thead>
<tbody>
<tr>
<td>Posterior parietal</td>
<td>Egocentric disorientation</td>
<td>Unable to represent the location of objects with respect to self</td>
<td>GW (Stark et al., 1997)</td>
</tr>
<tr>
<td>Posterior cingulate</td>
<td>Heading disorientation</td>
<td>Unable to represent direction of orientation with respect to external environment</td>
<td>Case 2 (Takahashi et al., 1997)</td>
</tr>
<tr>
<td>Lingual gyrus</td>
<td>Landmark agnosia</td>
<td>Unable to represent the appearance of salient environmental stimuli (landmarks)</td>
<td>AH (Pallis, 1955)</td>
</tr>
<tr>
<td>Parahippocampus</td>
<td>Anterograde disorientation</td>
<td>Unable to create new representations of environmental information</td>
<td>Case 1 (Habib and Sirigu, 1987)</td>
</tr>
</tbody>
</table>
of spatial representation that are clearly not unique to the representation of large scale, environmental space.

Except for Mr Smith, for whom lesion data are not available, all egocentrically disoriented patients described here had either bilateral or unilateral right lesions of the posterior parietal lobe, commonly involving the superior parietal lobe. Behavioural neurophysiology studies in monkeys and rodents tend to support the notion that cells within the posterior parietal cortex maintain representations of object position in an egocentric spatial frame. Studies of homologous cortical areas in monkeys have revealed cells with firing properties that represent the position of stimuli in both retinotopic and head centred co-ordinate spaces simultaneously (i.e. planar gain fields, Anderson et al., 1993). Notably, cells with exocentric firing properties have not been identified in the rodent parietal cortex, although cells responsive to complex conjunctions of stimulus egocentric position and egomotion have been reported (McNaughton et al., 1994).

In summary, there exists a group of patients who, following bilateral or right parietal lobe damage, display profound impairments in apprehending egocentric spatial relationships yet possess intact visual recognition abilities. One consequence of this basic spatial deficit is a general topographical disorientation that is likely the result of the inability to learn or recall appropriate spatial directions associated with (perhaps) properly perceived landmarks.

**Heading disorientation**

While the previous group of patients evidenced a global spatial disorientation, rooted in a fundamental disturbance of egocentric space, a second group of patients raises the intriguing possibility that exocentric spatial representations can be selectively damaged. A handful of patients have been described who are both able to recognize salient landmarks and do not have the dramatic egocentric disorientation described above. Instead, these patients are unable to derive directional information from landmarks that they recognize. It seems that they have lost a sense of egocentric direction, or ‘heading’ within their environment. Because there are so few of these cases, and because they have not been subjected to extensive testing, these observations must be regarded as tentative.

The three patients reported by Takahashi and colleagues are the best studied patients within this category (Takahashi et al., 1997). Case 2 in this study represents the group well: . . . as he was driving his taxi in the same city [in which he had worked for 6 years], he suddenly lost his understanding of the route to his destination. As he could quickly recognize the buildings and landscapes around him, he was able to determine his current location. However, he could not determine in which direction he should proceed. He stopped taking passengers and tried to return to the main office, but didn’t know the appropriate direction in which to drive.

Using the surrounding buildings, scenery, and road signs he made several mistakes along the way. He remembered, during this time, passing the same places over and over again.

All three describe being unable to derive directional information from the prominent landmarks that they recognize. These patients, however, present a rather different picture from those described as suffering from egocentric disorientation. Instead of being grossly disoriented in egocentric space, all three of the cases reported by Takahashi and colleagues were noted to have no signs of visuospatial agnosia (i.e. Balint’s syndrome or hemispatial neglect). Instead, it seems to be the case that these patients are unable to recall (or form) a link between directional information and landmark identity.

Takahashi and colleagues performed several tests with the three cases to confirm these observations. The assertion of the patients that they were still able to recognize landmarks was confirmed by a series of tests. The patients were able to ‘discriminate among buildings when several photographs were displayed’ and were able to recognize photographs of familiar buildings and landscapes near their homes. Also demonstrated to be preserved was the basic representation of egocentric space. The first and second cases of Takahashi and colleagues were tested for their memory of location of objects within a room: ‘. . . seven objects in the examination room (bookshelf, air conditioner, television, etc.) were pointed out and the patient was required to name them. Five minutes later both patients showed favorable recall of the positions of these objects . . . Moreover, both patients correctly recalled the names and positions of five objects that could be seen from the hospital window (warehouse, electrical power pole, etc.).’

Normal Corsi block performance was also demonstrated for patient 2. In contrast, performance on tests such as these was markedly impaired in the egocentric disorientation group.

In contrast to the preserved performance on these tests of landmark recognition and egocentric spatial memory, the patients of Takahashi and colleagues were impaired at the recall of previously learned topographical knowledge and impaired in the acquisition of novel information. For example, patients 1 and 2 were greatly impaired in map drawing tasks and all three were unable to describe routes between familiar locations and could not describe the positional (directional) relationship between one well-known place and another. In addition, the three patients were unable to draw a sketch-map of their hospital floor. It seems that these three cases are topographically disoriented but suffer neither from an inability to recognize landmarks nor from an inability to represent basic egocentric spatial relations. What, then, is the nature of their deficit?

Takahashi and colleagues suggested that the patients had lost a ‘sense of direction’ that allows one to recall the positional relationships between one’s current location and destination within a space that cannot be entirely surveyed at one time. This description captures the character of the
deficits reported in these patients, and is intriguing given the site of the lesion reported for these three patients: the right retrosplenial (i.e. posterior cingulate) region. Studies in rodents (Chen et al., 1994) have identified a small population of cells within this area that fire only when the rat is maintaining a certain heading, or orientation within the environment. These cells have been dubbed head-direction cells (Taube et al., 1996) and likely generate their signals based upon a combination of landmark, vestibular and idiothetic (self-motion) cues. Representation of the orientation of the body within a larger spatial scheme is a form of spatial representation that might be expected to be drawn upon for both route-based and map-based navigation, perhaps explaining the deficits observed in the patients reported by Takahashi and colleagues.

One other patient with topographical disorientation following a right cingulate lesion has been reported. Patient MB (Cammalleri et al., 1996) suffered from occasional bouts of ‘transient topographical amnesia,’ which the patient described as follows: ‘I was returning home when suddenly, even though I could recognize the places in which I was walking and the various shops and cafés on the street, I could not continue on my way because I no longer knew which way to go, whether to go forward or backward, or to turn to the right or to the left . . .’ Unfortunately, as MB was not examined during one of these events, only the patient’s description of the disorder is available.

The possibility of cingulate lesions producing isolated deficits in heading perception should be tempered, however, by the observation that difficulties with spatial orientation are not among the impairments typically reported in cases of ‘retrosplenial amnesia’ (Rudge and Warrington, 1991).

There has also been a report of transient (3 week) topographical disorientation following a right sided lesion of the dorsomedial and lateral posterior thalamus (patient OS, Kawahara et al., 1986). No description was given of the patient’s deficit, thus the case is mentioned here only as a curiosity, as head-direction cells have also been identified in the lateral dorsal thalamus of the rodent (Taube et al., 1996).

The possibility of isolated deficits in the representation of spatial heading is an intriguing one. These patients have a different constellation of deficits from those classified as egocentrically disoriented and the existence of these cases does suggest that separate cortical areas mediate different frames of spatial representation. Additional patients and more extensive testing are clearly needed, however, to better define the nature of this disorder.

Landmark agnosia
The third class of topographically disoriented patient can be described as landmark agnosic, in that the primary component of their impairment is an inability to use prominent, salient environmental features for the purposes of orientation. It should be noted at the outset that, despite the similarity in label, this category operates under a different set of assumptions from those of ‘topographical agnosia’ described by Paterson and Zangwill (Paterson and Zangwill, 1945). First, landmark agnosics do not generally have a deficit in perception of environmental information (i.e. object and spatial). Instead, they have a visual recognition deficit confined to salient environmental features. Secondly, landmark agnosics are not assumed to have perfectly intact perception. As has been described for the larger class of object agnosias (Farah, 1990), there is evidence that, when tested with sufficient rigour, associative agnosics display abnormal perceptual skills. Finally, as both perceptual and mnestic substrates are proposed to be damaged in landmark agnosics (as they are one and the same), these patients display disorientation in both novel and previously familiar environments.

There are several well-studied cases within this category, including patients J.C. (Whiteley and Warrington, 1978), A.R. (Hécaen et al., 1980), SE (McCarthy et al., 1996), M.S. (Incisa della Rocchetta et al., 1996), several of the cases reported by Landis and colleagues (Landis et al., 1986), the cases reported by Takahashi and colleagues (Takahashi et al., 1989), Funakawa and colleagues (Funakawa et al., 1994) and Suzuki and colleagues (Suzuki et al., 1996). One of the earliest and most comprehensively described reports was that of Pallis (Pallis, 1955) concerning patient A.H.:

He complained a lot of his inability to recognize places. ‘In my mind’s eye I know exactly where places are, what they look like. I can visualize T . . . Square without difficulty, and the streets that come into it . . . I can draw you a plan of the roads from Cardiff to the Rhondda Valley . . . It’s when I’m out that the trouble starts. My reason tells me I must be in a certain place and yet I don’t recognize it. It all has to be worked out each time . . . For instance, one night, having taken the wrong turning, I was going to call for my drink at the Post Office.’ He seemed to have difficulty in assimilating new topographical data. ‘It’s not only the places I knew before all this happened that I can’t remember. Take me to a new place now and tomorrow I couldn’t get there myself.’ His topographical memory was good, as could be inferred from his accurate descriptions of paths, roads, the layout of the mine-shafts [the patient was an engineer], and from his excellent performance in drawing maps of places familiar to him before his illness. There was no evidence of neglect or imperception of any part of extra-personal space, localization of objects in space was excellent . . . He would have difficulty in reconciling the reality about him with the plan in his mind in convincing himself that he was in a given situation. ‘I have to keep the idea of the route in my head the whole time, and count the turnings, as if I were following instructions that had been memorized.’ He could at a glance tell terraced council-houses from detached villas, a living room from an office, a country lane from a main road.
There are several salient features of this description to note. First, A.H. was disoriented both in previously familiar environments and novel places. Secondly, there is the suggestion that his manipulation of spatial information per se, as judged by his intact map production, was intact. Thirdly, despite being able to distinguish different classes of buildings, patient A.H. was unable to identify specific buildings. These three features are common to all of the cases listed above. Despite a preserved ability to provide spatial information regarding a familiar environment, the patient is unable to way-find because of his inability to recognize prominent landmarks.

This loss of landmark recognition, and its relative specificity, has been formally tested by several authors, usually by asking the subject to identify pictures of famous buildings. Patient S.E. (McCarthy et al., 1996) was found to be markedly impaired at recalling the name or information about pictures of famous landmarks and buildings compared with both the performance of control subjects and his performance with recalling information regarding famous people. Patient M.S. (Incisa della Rocchetta et al., 1996) performed at chance on three different delayed recognition memory tests that used pictures of (i) complex city scenes, (ii) previously unfamiliar buildings, and (iii) country scenes. M.S. was also found to be impaired at recognizing pre-morbidly familiar London landmarks. Takahashi and colleagues (Takahashi et al., 1989) obtained 17 pictures of the patient’s home and neighbourhood. The patient was unable to recognize any of these, but he could describe from memory the trees planted in the garden, the pattern printed on his fence, the shape of his mail box and windows and was able to produce an accurate map of his house and home-town.

In contrast, tests of spatial representation have generally shown intact abilities in these patients. Patients S.E. (McCarthy et al., 1996), M.S. (Incisa della Rocchetta et al., 1996) and J.C. (Whiteley and Warrington, 1978) were all found to have normal performance on a battery of spatial learning and perceptual tasks that included Corsi span, Corsi supra-span and ‘stepping-stone’ mazes (Milner, 1965). [Patient A.R. (Hécaen et al., 1980), however, was found to be impaired on the last of these tests.] In general, the ability to describe routes and produce sketch-maps of familiar places is intact in these patients. As was discussed previously, these more anecdotal measures of intact spatial representation should be treated with some caution as there is considerable ambiguity as to the specific nature of the cognitive requirements of these tasks. Nonetheless, the perfectly preserved ability of patients A.R. and A.H. (Pallis, 1955) to provide detailed route descriptions, and the detailed and accurate maps produced by S.E., A.H. and the patient of Takahashi and colleagues (Takahashi et al., 1989), are suggestive of intact spatial representations of some kind. (Patient M.S., however, was noted to have poor route description abilities.) Moreover, particularly compelling are reports of patients producing accurate maps of places that were not familiar prior to the lesion event (Cole and Perez-Cruet, 1964; Whiteley and Warrington, 1978). In this case, the patient can only be drawing upon preserved spatial representational abilities to successfully transform navigational experiences into an exocentric representation.

Several neuropsychological deficits have been noted to co-occur with landmark agnosia, specifically, prosopagnosia (Pallis, 1955; Cole and Perez-Cruet, 1964; Landis et al., 1986; Takahashi et al., 1989; McCarthy et al., 1996) and achromatopsia (Pallis, 1955; Landis et al., 1986), along with some degree of visual field deficit. These impairments do not invariably accompany landmark agnosia (e.g. Hécaen et al., 1980), however, and are known to occur without accompanying topographical disorientation (e.g. Tohgi et al., 1994). Thus, it is unlikely that these ancillary impairments are actually the causative factor of topographical disorientation. More likely, the lesion site that produces landmark agnosia is close to, but distinct from, the lesion sites responsible for prosopagnosia and achromatopsia.

According to the classic ‘associative agnosia’ model, patients should have normal perception of environmental features that are devoid of meaning. Only two of the landmark agnosics have been examined in detail with regard to their perceptual abilities. Patient A.R. (Hécaen et al., 1980) was able to perform a ‘cathedral matching’ task accurately, but: ‘... he [A.R.] spontaneously indicated that he was looking only for specific details ‘a window, a doorway... but not the whole’. . . Places were identified by a laborious process of elimination based on small details.’

Patient J.C. (Whiteley and Warrington, 1978) was administered a building matching task that he performed perfectly, but no details are given regarding the character of his performance and, notably, control subjects also scored near ceiling. Further studies of the perceptual abilities of these patients will be necessary to settle the question. Finally, patient S.E. (McCarthy et al., 1996) was noted to have a loss of semantic knowledge regarding buildings in addition to the visual impairments. As no other landmark agnosic has been tested for landmark semantic knowledge, it is not possible to state whether semantic loss is common to landmark agnosic patients or peculiar to S.E. Again, additional studies will be necessary.

An additional hallmark feature of landmark agnosia is the compensatory strategy employed by these patients. The description of patient J.C. (Whiteley and Warrington, 1978) is typical: ‘He relies heavily on street names, station names, and house numbers. For example, he knows that to get to the shops he has to turn right at the traffic lights and then left at the cinema . . . When he changes his place of work he draws a plan of the route to work and a plan of the interior of the ‘new’ building. He relies on these maps and plans . . . He recognizes his own house by the number or by his car when parked at the door.’

This reliance upon small environmental details, termed variously ‘signs’, ‘symbols’ and ‘landmarks’ by the different authors, is common to all of the landmark agnosia cases described here and provides some insight into the cognitive
nature of the impairment. First, it is clear that these patients are capable of representing the strictly spatial aspect of their position in the environment. In order to make use of these minute environmental details for way-finding, the patient must be able to associate spatial information (if only left or right turns) with particular way points. This is again suggestive evidence of intact spatial abilities. Secondly, although these patients are termed landmark agnosics, it is not the case that they are unable to make use of any environmental object with orienting value. Instead, they seem specifically impaired in the use of high salience environmental features, such as buildings, and the arrangement of natural and artificial stimuli into scenes. Indeed, these patients become disoriented within buildings, suggesting that they are no longer able to represent the configuration of stimuli which suffices to differentiate easily one place from another. It thus seems that careful study of landmark agnosics may provide considerable insight into the normative process of selection and utilization of landmarks.

A possible account of landmark agnosia, then, posits that there exists a cortical region that is predisposed to the representation of visual information that is engaged in way-finding. Through experience, this area comes to represent environmental features and visual configurations which have landmark value (i.e. tend to aid navigation). We might imagine such a spatially segregated, specialized area would develop because of the natural correlation of landmark features with other landmark features (Polk and Farah, 1995). Lesions to this area would produce the pattern of deficits seen in the reported cases of landmark agnosia. It should be noted that this model of landmark agnosia explicitly eschews the notion that landmark-based topographical disorientation is simply ‘more of the same’ impairment that produces general object agnosia or prosopagnosia. Instead, the proposal made here is that there exists a specialized neuroanatomical substrate for the representation of topographical information, distinct from other ‘ventral stream’ areas involved in, for example, face perception.

Interestingly, the lesion sites reported to produce landmark agnosia are fairly well clustered. Except for patient J.C. (Whiteley and Warrington, 1978) (who suffered a closed head injury and for whom no imaging data are available) and patient M.S. (Incisa della Rocchetta et al., 1996) (who suffered diffuse small vessel ischaemic disease), the cases of landmark agnosia reviewed here all have lesions either bilaterally or on the right side of the medial aspect of the occipital lobe, involving the fusiform and lingual gyri and sometimes the parahippocampal gyrus. The most common mechanism of injury is an infarction of the right posterior cerebral artery.

Recent neuroimaging studies have tested some aspects of this proposed organization of ventral cortex. Using functional MRI, we have identified a cortical area that responds with a greater magnitude of neural responses to buildings compared with other stimuli, including faces, cars, general objects and phase-randomized buildings (Aguirre et al., 1998a). Across subjects, the voxels that evidenced ‘building’ responses were located straddling the anterior end of the right lingual sulcus, just posterior to the parahippocampus. This site and its responses are in good agreement with the predictions of the landmark agnosia model presented here. The finding of ‘building sensitive’ cortex within the anterior, right lingual gyrus has been replicated by other groups (Haxby et al., 1999).

In summary, the evidence for the existence of a cortical region specialized for the representation of topographical information seems strongest in the case of the landmark agnosics. These patients are among the best studied cases of topographical disorientation, and seemingly the most numerous. Further examination of the deficits evidenced by these patients may be very informative as to the organization of visual systems involved in the selection and utilization of salient environmental landmarks.

**Anterograde disorientation**

All of the cases of topographical disorientation described to this point have evidenced some degree of impairment both in previously familiar and novel environments. This finding is consistent with a model of cortical memory which suggests that the same neural substrates mediate the perception and long-term storage of particular types of information (Squire and Zola, 1996). Cases have been reported, however, of topographical impairment primarily confined to novel environments. Interestingly, this anterograde disorientation, described in two patients by Ross (Ross, 1980), one patient by Pai (Pai, 1997) and the first two cases reported by Habib and Sirigu (Habib and Sirigu, 1987), appears to impact both landmark and spatial spheres. The following description is from the first case reported by Ross:

The major problem he noted, besides a complete inability to recognize faces, was severe spatial disorientation. In order to find his way around the college campus or to walk to and from school, he was constantly forced to consult maps and written notes . . . The patient had no difficulty in accurately reaching for objects in space . . . The patient had no difficulty in moving through his environs. He did not appear or act as though he were blind. Nevertheless, he was never able to learn the spatial organization of the neurology wing during the entire month he spent in the hospital . . . When asked to construct a map of the neurology wing, he was able to do this task if allowed to walk through the ward but was unable to do it from memory . . . the patient also appeared spatially disoriented in the three-room apartment where he had been living for six months. In striking contrast, however, when he stayed at his parents’ house, in which he grew up, there was no observable difficulty with spatial orientation.

All four patients displayed preserved way-finding in environments known at least 6 months before their lesion. Patient 1 in the study by Ross was able to draw a very accurate map of his parents’ home, and both case 1 and case
2 in the study by Habib and Sirigu reported that, following an initial period of general impairment, no orientation difficulties were encountered in familiar parts of town. These patients were tested for recognition of photos of familiar places and buildings and succeeded globally, although with some latency. Also, these patients do not demonstrate gross spatial disorientation, in the manner of patients G.W. (Stark et al., 1996) or M.N.N. (Kase et al., 1977). Instead, the spatial component of their disorder is primarily manifest as an inability to produce maps of novel environments from memory, although patient 1 in the Ross study was also impaired in remembering the location of objects in egocentric space across a delay.

In novel environments, however, these patients encountered great difficulty. In the Habib and Sirigu study, case 1 was unable to learn a route within the hospital corridor: ‘when he had to go from the psychologist’s office back to his room, he wandered along the ward, being unable to find his room unless he relied on the sequence of door numbers’ and ‘when asked to draw a floor plan of this simple path, he invariably produced an erroneous drawing, omitting several turns making up the path and misplacing the office in relation to the main landmark (the elevators).’ Case 2 reported that she ‘had to pay attention to verbal cues such as inscriptions on store windows or street names.’ The second case reported by Ross remained disoriented even after 6 days in the neurology ward where he was staying.

Ross attributed the impairments found in his patients to a specific loss of recent visual memory. His patients were able to learn and recall information conveyed by tactile and auditory channels without difficulty. However, if a patient was shown a relatively non-verbalizable object, he could not select it from among four similar objects several minutes later. In agreement with these findings, the patients reported by Habib and Sirigu were found to have impaired visual compared with verbal memory on the Wechsler Memory Scale and performed very poorly on the recall from memory of Rey–Osterreith’s figure.

In each of these cases, the lesion site has been within inferior ventral cortex on the right side. This general location is similar to that reported for landmark agnosia, but, at least in the cases of Habib and Sirigu and Pai (Pai, 1997), more precise localization to the parahippocampus can be made. As there are several similarities between this category of deficit and that of the landmark agnosics, it remains to be conclusively demonstrated that they constitute two separate entities, instead of different manifestations of lesions to a common system. However, the preponderance of anterograde deficits in this group does suggest that the critical site of damage may be within the medial temporal lobe (i.e. parahippocampus), given the known involvement of this area in declarative memory formation (Squire and Zola, 1996).

Recent neuropsychological reports have examined the effects of lesions within the parahippocampus. Twenty patients with unilateral medial temporal lesions (half on either side) were tested on a video-taped route learning task (Maguire et al., 1996a). While these patients denied retrograde topographical disorientation and did not have any measurable general memory impairments, they were impaired relative to controls on tests of route learning and exocentric position judgements. All 20 patients had surgical resections of the parahippocampal gyrus. The authors did not comment upon any difference in performance or clinical presentation between the patient groups with small (excluding hippocampus) or large (including hippocampus) resections. Interestingly, patients with left or right excisions had roughly equivalent impairments. Another report (Bohbot et al., 1998) also suggests the involvement of the parahippocampus in topographical learning, although it found different effects of laterality. Fourteen patients with well defined thermo-coagulation lesions of the medial temporal lobes were tested on a human analogue of the Morris water maze task (Morris et al., 1982). Patients with lesions confined to the right parahippocampal cortex were impaired relative to those with lesions of the left parahippocampal cortex, right or left hippocampus and epileptic controls.

Thus, there is evidence that the parahippocampus subserves some computational function necessary for the acquisition of novel topographical knowledge. A specialized role in spatial learning (broadly defined) has been proposed previously for the parahippocampal cortex, based upon the neocortical inputs to the area (including inferior caudal visual areas, retrosplenial cortex and the superior parietal lobule) (Suzuki and Amaral, 1994). As has been described above, lesions to these other cortical areas also produce different varieties of disorientation. Thus, the parahippocampal cortex is in a position to associate particular landmarks (represented in ventral occipitotemporal regions) with particular spatial relationships (represented in posterior parietal and retrosplenial cortex) (McNaughton et al., 1989).

Of particular interest are recent neurophysiological studies of the parahippocampal area in freely moving monkeys. Rolls and colleagues (Rolls et al., 1997) have identified ‘spatial view’ cells which respond when the monkey looks at a particular part of the environment. The authors have suggested that a population of these cells might be used to encode the location of particular objects in space or to build representations of exocentric location within a detailed environment. A number of neuroimaging studies in humans that have attempted to isolate ‘topographical’ cognitive processes have also identified activity within the parahippocampus (Aguirre et al., 1996; Maguire et al., 1996b, 1997, 1998a). While intriguing, these studies uniformly suffer from considerable limitations in inferential power due to the unsupported assumptions of ‘cognitive subtraction’ (Friston et al., 1996) under which they were conducted. As a result, they can only be taken as suggestive of parahippocampal involvement in topographical processes in the human (for a critique of these investigations, see Aguirre et al., 1998b).

A recent study by Epstein and Kanwisher has, however, provided more inferentially sound observations regarding the functional activity of the parahippocampus (Epstein and
Kanwisher, 1998). Evoked activity within the parahippocampus was found to be much greater in response to the presentation of ‘places’—pictures that represent a spatially extended location, such as rooms and outdoor scenes. Dramatically, these responses were equivalent whether the pictured rooms contained any objects or simply bare walls! These findings are particularly interesting when considered in light of the ‘view cells’ identified in the monkey. Evidence from several different methodologies, therefore, now argues for a role for the parahippocampus in representing topographical information.

The hippocampus
Any discussion of knowledge of place would be incomplete without mention of the hippocampus. This paleocortical area has held special significance within the study of environmental representation following description of ‘place-cells’ within the rat hippocampus that fire preferentially when the animal is in a specific location within its environment (O’Keefe and Dostrovsky, 1971). This finding led to the proposal that the hippocampus maintains a ‘cognitive map’ of learned environments, with the pattern of activity indicating the current position within that space (O’Keefe and Nadel, 1978). Additional evidence regarding the importance of the hippocampus in topographic learning was provided by the report of Morris and colleagues (Morris et al., 1982) that rats with hippocampal lesions were impaired on a specific test of place learning—the water maze task. The specificity of the role played by the hippocampus (i.e. Ammon’s horn, the dentate gyrus and the subiculum) in spatial representation has subsequently been debated at length (e.g. Cohen and Eichenbaum, 1993). At the very least, it is clear that selective (neurotoxic), bilateral lesions of this structure in the rodent greatly impair performance on ‘place’ learning tasks such as the water maze (Morris et al., 1990; Jarrard, 1993).

The importance of the hippocampus for exocentric spatial representation in the human has been more difficult to demonstrate. Spatial memory tests that present fixed stimulus arrays to a stationary patient (e.g. Smith and Milner, 1981; Cave and Squire, 1991) are not strictly relevant, as the cognitive map theory proposes a flexible, map-like representational role for the hippocampus. Interestingly, unilateral lesions of the hippocampus have not been reported to produce any appreciable real-world way-finding impairments in humans (De Renzi, 1982). The existence of anterograde way-finding deficits in patients with general anterograde amnesia following bilateral lesions of the hippocampus (and adjacent structures) (Scoville and Milner, 1957; Zola-Morgan et al., 1986; Rempel-Clower et al., 1996) has not been explicitly commented upon. However, if present, any topographical difficulties would obviously be accompanied by memory impairments in other areas. Retrograde loss of way-finding knowledge in these patients is not apparently disproportionate to losses in other areas (Rempel-Clower et al., 1996) and can be preserved (Milner et al., 1968). Based upon these findings, if the hippocampus is indeed necessary for the representation of topographical space in humans, then it must be said (i) that either hemisphere is sufficient to support this function, (ii) that way-finding in previously learned places can be accomplished in its absence, and (iii) that place learning is but one of many kinds of knowledge for which it is necessary (i.e. place learning is a type of declarative memory) (Squire and Zola, 1996).

Conclusions and future directions
We have attempted to provide a framing taxonomy for the collected cases of topographical disorientation. Some portions of this taxonomy, particularly the designation of egocentric disorientation and landmark agnosia, seem well supported by the neuropsychological evidence and are concordant with the results of electrophysiological and functional imaging studies. Other components, however, remain tentative designations in need of corroborating evidence from future neuropsychological and imaging studies. In particular, several outstanding questions remain. For the cases of landmark agnosia, it remains unclear what type of visual information is represented at the critical lesion site. Is this a region involved in the representation of all ‘landmark’ information, or simply certain object classes that happen to be used as landmarks? Careful testing of the recognition abilities of patients who suffer from this disorder will better define the representational responsibilities of this cortical area and may inform as to the normative selection of visual objects for use as landmarks.

Is anterograde disorientation a distinct disorder? The parahippocampal lesion site we have associated with this classification is very similar in location to that identified for landmark agnosia. Additionally, neuroimaging studies have activated both parahippocampal and anterior lingual sites in association with topographical representation. These cortical areas are closely apposed and, in fact, no clear anatomical boundary is present to distinguish them. Future studies will be required to determine if there are indeed separate cortical sites associated with anterograde disorientation and landmark agnosia, and if the functional behaviour of these regions differs.

The breadth and heterogeneity of the topographical disorientation literature is at once both a burden and an opportunity. While the complexity of the behaviours under study makes tidy summary of the cases challenging, topographical disorientation provides a window into the integration and organization of several interesting cognitive processes. Those, whose interests encompass object recognition, spatial representation, memory or any one of several other fields, might find insight into these basic cognitive processes through consideration of the cases examined here.

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