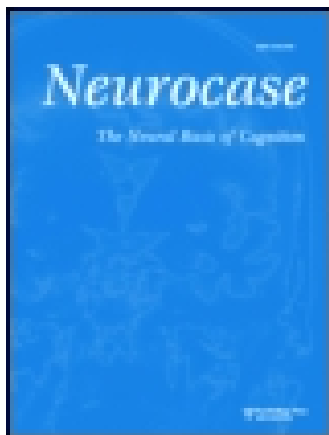


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Preserved Knowledge of Maps of Countries: Implications for the Organization of Semantic Memory

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Preserved Knowledge of Maps of Countries: Implications for the Organization of Semantic Memory

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Abstract

We describe two patients with selectively preserved knowledge of the category of countries. Following a series of cerebral infarcts, patient DB presented with severe perceptual impairment, including dense apperceptive agnosia, prosopagnosia, and topographical agnosia. Despite these deficits, he could effortlessly name countries from their outline maps. Patient WH, who suffered from semantic dementia, had severe naming and comprehension difficulties, with extremely sparse residual semantic knowledge. Remarkably, the category of countries was preserved. First, we argue that, for both patients, this category preservation occurs at a semantic level. Second, we discuss our findings in the context of three current models of category-specific effects (perceptual, ontogenetic, and evolutionary models). We argue that the perceptual model (Humphreys and colleagues) cannot easily accommodate our findings. By contrast, the ontogenetic (Warrington and colleagues) and evolutionary models (Caramazza and colleagues) can explain our findings. However, some modifications to both models are required. The ontogenetic model needs to envisage a spatial channel for the development of map knowledge, which is anatomically separate from channels of other categories of knowledge. The evolutionary model needs to envisage the possibility that some categories of knowledge, such as countries, may not be prewired, but learned during ontogenetic development.

Introduction

Category-specific deficits of semantic memory can be highly selective (see Gainotti, 2000 for a review), as, for example, in the category of proper nouns. Semenza and Zettin (1988, 1989) first reported two patients who were impaired in retrieving proper names (famous people as well as geographical names, such as cities, rivers, countries, and mountains) despite being able to retrieve common names. Similar findings were reported thereafter (e.g., Harris and Kay, 1995; Papagno and Capitani, 1996). There are also reports of yet more selective proper noun deficits, limited to people's names (e.g., Hittmair-Delazer *et al.*, 1994; Fery *et al.*, 1995; Miceli *et al.*, 2000).

The reverse dissociation, selective *sparing* of proper nouns is quite rare, and only a few examples have been documented thus far (Cipolotti *et al.*, 1993; Semenza and Sgaramella, 1993). Semenza and Sgaramella described a patient whose only intelligible output in spontaneous speech was people's names. On confrontation naming, he could only retrieve people's names, although only with phonemic cueing. The patient

reported by Cipolotti *et al.* showed intact written naming for famous people (e.g., Virginia Woolf) and country names (e.g., Scotland). In sharp contrast, her written naming for common names was abolished, even for very familiar items (e.g., chair).

Selective sparing of a specific category of proper nouns has also been described: that of the category of countries. Patients with preserved ability to retrieve country names have been reported (e.g., McKenna and Warrington, 1978; Warrington and Clegg, 1993). Incisa della Rocchetta *et al.* (1998) described a series of patients who, despite being virtually at ceiling in retrieving country names, were severely impaired in retrieving common names. Interestingly, in one of these patients (TM), country names were selectively preserved not only with respect to common names, but also with respect to famous people and building names. Ward *et al.* (2000) recently documented another patient in whom country names were selectively preserved despite a severe impairment in producing not only common names but also famous people's

names. Moreover, a modality-specific effect for the names of countries has been documented at the level of lexical output. Cipolotti (2000) described a patient (BF) with a striking modality-specific selective sparing of country and nationality names with concurrent, severe impairment of common nouns and famous people. In the oral output modality (oral naming and reading aloud), BF was severely impaired in producing a wide variety of common and proper nouns, except for country and nationality names, such as *France* and *Spanish*. In the written output modality (written naming and writing to dictation) she no longer had difficulties with common nouns, but was again impaired on proper nouns, except for country names.

This evidence of selective preservation of country knowledge and country names has been interpreted as compatible with the notion that countries are processed in brain regions that are information-encapsulated and distinct from those involved in processing other such categories of proper and common nouns (Cipolotti, 2000).

In more general terms, these neuropsychological studies of patients with category-specific effects have offered new insights into the organization of the semantic system. In this context, it should be noted that at present there is not a general consensus with regard to the theoretical underpinnings of category-specific effects. The most commonly held views fall into three camps. The first view holds that category effects occur outside the semantic system. The other two views propose that the effects occur within the semantic system. However, they diverge with respect to the organisation of knowledge inside the semantic system itself.

The first view considers the locus of category-specific effects to occur at a perceptual level. Its basic postulate holds first that within-category errors are due to competition between perceptually similar items at early levels of visual processing. Errors will propagate forward to subsequent levels in *cascade* fashion and thus spread to the semantic and phonological systems (Humphreys *et al.*, 1988, 1997; see also Caramazza *et al.*, 1990; Gaffan and Heywood, 1993; Rapp *et al.*, 1993; Hillis *et al.*, 1995). Categories with many perceptual competitors (such as living things) are more prone than categories with fewer competitors to this kind of error. In this view, a category-specific impairment which appears to affect the semantic level can be caused instead by a deficit in a presemantic level. Recent neural network simulations have given some support for this hypothesis (Gale *et al.*, 2001). Although this model was elaborated to deal with the living/nonliving dichotomy, its assumptions and predictions could be easily generalized to any category whose exemplars are visually similar, such as faces, buildings, and maps. The model implies that patients with visual perceptual deficits are likely to develop category-specific deficits involving categories with visually similar exemplars.

The second view considered category-specific naming impairment to occur at the semantic level (e.g., Warrington and Shallice, 1984; Basso *et al.*, 1988; De Renzi and Lucchelli, 1994; Barbarotto *et al.*, 1995; Caramazza and

Shelton, 1998). In this framework, the selectivity of deficits would reflect selective impairment of one of two distinct subsystems, one encoding the sensory features of objects, the other processing their functional characteristics (Warrington and Shallice, 1984). In an update of this position, McCarthy and Warrington (1987, 1990) argued that semantic categories are shaped during ontogenesis. The way in which they are acquired in childhood would determine how they are encoded and stored by the brain. Thus, a given category may rely more on certain channels of information than another category (e.g., sensory channels would be preferred for animals, whereas manipulable objects would preferentially engage motor channels).

The third view is held by Caramazza and Shelton (1998), with the hypothesis that fractionations are shaped during phylogenesis. Evolutionary pressures would result in specialized neural mechanisms devoted to distinguishing *perceptually* and *conceptually* specific kinds of objects, leading to the categorical organization of knowledge in the brain. However, as Caramazza (2000, p. 1043) pointed out "... The range of categories that can be subsumed under this principle is very narrow – it is restricted to categories of objects whose rapid and accurate classification would have led to survival and reproductive advantages. Plausible candidates are the categories of conspecifics, animals and plant life (and perhaps artifacts [sic])."

In this article we present two patients showing selective sparing of the category of countries. Patient DB presented with severe visuo-perceptual deficits. Despite these, he had no difficulty whatsoever at identifying countries from outline maps. However, he was unable to recognize faces and buildings. Patient WH showed a profound and severe impairment of semantic knowledge for a large number of common and proper noun categories. By contrast, her ability to comprehend map outlines was spared. We argue that these findings challenge the perceptual hypothesis advanced by Humphreys and collaborators (Humphreys *et al.*, 1988, 1997; Gaffan and Heywood, 1993) and require some modification of both the ontogenetic view of Warrington and colleagues and the evolutionary perspective proposed by Caramazza and Shelton (1998). We will argue that the ontogenetic view needs to envisage a spatial channel for the development of map knowledge, which is anatomically separate from those of other categories of knowledge. Its representation would thus be anatomically separate from that of other categories of knowledge that rely on other sensory channels, such as shape and texture. Caramazza and Shelton's hypothesis needs instead to allow for sufficient flexibility in the nervous system to code for novel categories.

Case 1

Patient DB was a 67 year-old former electronics analyst. In June 2000 he noted unsteadiness and lurching when walking and was later found collapsed in the street. He was admitted to the National Hospital for Neurology and Neurosurgery under

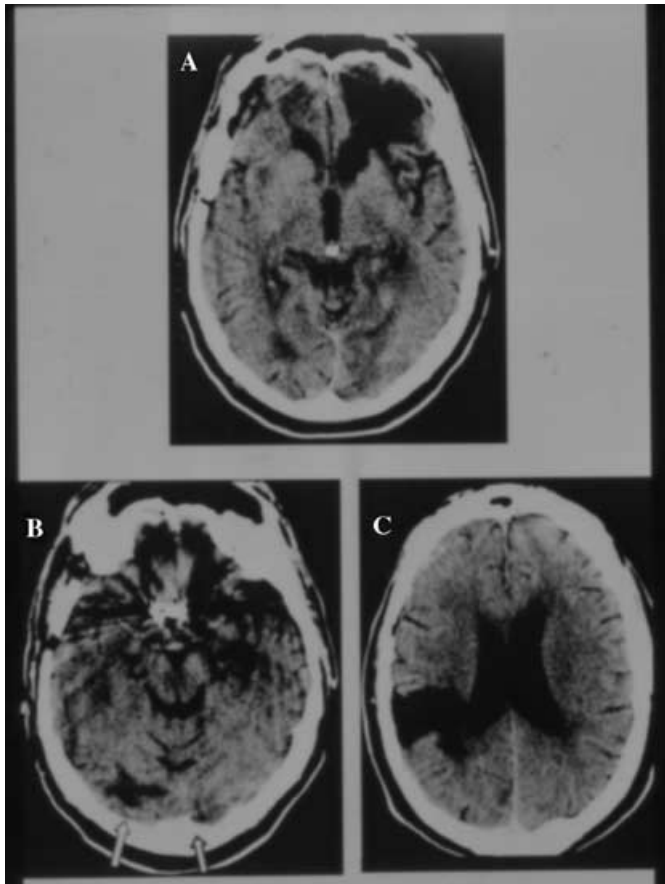


Fig. 1. CT scan images of Patient DB: A. Left frontal infarct. B. Bilateral occipital infarcts (indicated by arrows). The infarct on the right is appreciably larger than the one on the left. The right parahippocampal gyrus appears intact. C. Right parietal infarct. It appears to involve the anterolateral aspect of neocortex and underlying white matter, but spare the more dorsolateral aspect. Note that the occipital infarct visible in B is no longer discernible at this level. This suggests sparing of the superior aspect of the occipital lobe.

the care of Dr. Peter Rudge. His past medical history included hypertension and neurosurgical intervention for clipping of an anterior communicating artery aneurysm (in 1993). Neurological examination demonstrated a left homonymous hemianopia with incomplete right upper quadrantanopia and macular sparing. Sluggish ankle jerk reflexes and blunting of vibration sense in the left foot were noted together with mild unsteadiness on tandem walking. CT scans revealed an old infarct in the left frontal region (Fig. 1A). New infarcts were noted in the occipital regions bilaterally (more marked on the right; Fig. 1B), and in the right parietal area (1C). The aetiology of the infarcts was uncertain, the most likely being arthero-sclerotic.

Neuropsychological assessment

DB was referred to the Neuropsychology Department, where he underwent formal neuropsychological assessment. Table 1 shows his baseline cognitive scores. His WAIS-R scores are both in the average range, which is only slightly below his

Table 1. Patient DB. Baseline cognitive scores

<i>General Intelligence</i>		
VIQ	101	
PIQ	93	
NART IQ	110	
<i>Memory</i>		
Recognition Memory Test		
Words	18/25	<5 th %ile
Faces	14/25	<5 th %ile
<i>Executive Functions</i>		
Weigl Sorting Test	2 Categories	
Proverbs	3/4	
<i>Language</i>		
Graded Naming Test		
Objects	13/30	at 5 th %ile
Proper Nouns	9/30	50 th %ile
Naming by definition	9/15	<5% cut off (12/15)
Naming Famous Faces	3/12	<5 th %ile
Naming Maps	10/10	
<i>Perception</i>		
Visuoperceptual		
Object Decision*	7/20	<5% cut off (14/20)
Silhouettes*	6/30	<5% cut off (15/30)
Unusual Views	2/20	<5% cut off (13/20)
Usual Views	16/20	<5% cut off (18/20)
Same/different faces	13/20	<5% cut off (15/20)
Same/different buildings	10/20	Weak (Lowest control score = 15/20)
Incomplete Letters*		
Shape Detection*	11/20	<5% cut off (16/20)
Satisfactory: 5% cut off = (15/20)		
Visuospatial		
Number Location*	10/10	
Position Discrimination*	20/20	
Dot Counting*	10/10	

*VOSP Subtests.
%ile = percentile.

high-average premorbid optimal level of functioning, as estimated by his reading on the National Adult Reading Test (NART; Nelson, 1982). He scored very poorly on the short Recognition Memory Test (Warrington, 1996). He passed the Weigl sorting test, a simple test of frontal-executive functioning, but his performance was slightly weak in a proverb interpretation task. He had mild word retrieval difficulties on naming tests involving common nouns (Common Noun Graded Naming Test: McKenna and Warrington, 1983; Naming by Definition: Coughlan and Warrington, 1978), but he performed within the average range on proper nouns (Proper Noun Graded Naming Test: McKenna and Warrington, 1983) and scored almost flawlessly (48/50) in naming high-frequency pictures from a naming test comprising 5 categories (Maps, Animals, Body Parts, Objects and Colours – see below: Patient WH, Experimental Investigation, Task 1).

There was evidence of prosopagnosia. DB had a severe deficit in naming and identifying famous people from photographs (Warrington and James, 1967). By contrast, when given their names he could readily identify them, by describing their professional background (e.g., Who is John Major: “He is an ex-Prime Minister.”). Despite this severe deficit, his ability to

name maps of countries was intact: remarkably, DB was able to name correctly 10 maps of European countries.

DB had marked visual perceptual difficulties. We assessed his visual perception with four subtests from the Visual Object and Space Perception Battery (VOSP; Warrington and James, 1991), and with the Unusual Views Test (Warrington and Taylor, 1973; see also: Warrington and James, 1988). In the Object Decision and Silhouettes tests DB had to identify objects from foreshortened silhouettes. His performance was markedly impaired on both tasks. Likewise, he found objects photographed from unusual perspectives almost impossible to identify (Unusual Views). However, he had only very mild difficulty when the photographs were taken from conventional perspective (Usual Views). His performance was poor in making same/different judgements of faces and buildings in tasks in which the stimuli to be matched were photographed from different perspectives (Whitely and Warrington, 1978; Warrington and James, 1988). He performed poorly in identifying fragmented letters. Despite this, his performance was only mildly weak on a test of early visual processing (Shape Detection).

In sharp contrast to his impaired visuoperceptual functions, DB's visuospatial skills were well preserved. He scored flawlessly on three subtests from the VOSP. These all required spatially based judgements, such as identifying the spatial location of a dot within a frame (Number Location), deciding which of two dots was at the centre of a square frame (Position Discrimination), or counting scattered dots (Dot Counting).

Summary. DB presented with prosopagnosia and marked apperceptive agnosia. Despite these profound deficits, he was still able to name maps. His spatial perception was intact.

Experimental investigations

The puzzling finding that emerged from the neuropsychological assessment was that, despite his profound prosopagnosia and apperceptive agnosia, DB had no difficulty at naming maps. These aspects of his performance were the focal point of interest of the experimental investigations.

In the following experimental investigation, we examine in more detail DB's residual perceptual skills and map knowledge. In testing visuoperceptual skills we attempted to evaluate two types of processes: the ability to access the internal representations of objects and the ability to identify objects under conditions of stimulus degradation.

Testing DB's residual perceptual skills; Internal representation of objects

Task 1: Verbal description of familiar objects and famous faces

Control participants. Five normal-control participants of similar background to DB, but slightly younger (average age: 59), were enrolled in the tasks described below.

Material, procedure and scoring. We first asked DB to provide a verbal description for two categories of items:

familiar objects (e.g., train, binoculars; $n = 15$) and faces of famous people (e.g., Sean Connery, Margaret Thatcher; $n = 15$). The control participants were asked to provide verbal descriptions of the same items with instructions identical to those used with DB. Independently of DB and of the controls, we generated a list of features for each of the objects and for the faces (with the help of photographs). For example, for the bicycle, our list included: frame, wheels, spokes, pedals, chain, handle bars and saddle. For Princess Diana, our list consisted of the following: Age, 20s–30s; Hair color, blond; face, square/oval; eyes, blue; nose, long/bump/crooked; mouth, full; body, tall and slim; glasses, no. We then compared the list generated by us to the list of features generated by the controls and produced a template protocol (which we used for scoring) by adding to our list any feature generated by the controls (but not DB) that we had not included in our original list. The template protocol thus resulted in a total of 96 features for the 15 objects and 103 for the 15 faces. We excluded functional features (*Container* for Bottle, *singer* for Elvis) from the template protocol, even if produced by the control participants. Likewise, we excluded arbitrary features such as *sexy* or *voice of an angel*. Two independent raters, blind to the identity of the participants and to the purpose of the experiment, scored each individual participant's protocol (DB and 5 controls) by tallying the features they produced against the lists of features in the template protocol. They awarded one point for each feature. Since there was a high correlation between the two raters ($r = 0.95$), the scores we used in the analysis were the average of the two raters' score.

For the routes task, DB was asked to describe verbally seven routes from his house to familiar neighboring locations (e.g., Underground station, GP's office). Six control subjects were recruited from the first author's neighborhood (they lived all in very close proximity) and asked to describe routes to familiar neighboring locations, as for DB. The routes used with the controls were all, as for DB, within 1 km² of their houses. They were chosen such that they would be equivalent to DB's routes in distance, number of turns and salience of the destinations (Underground station, GP's office). One point was given for each correct turn and intersection mentioned. The scoring of routes was given as a percentage correct since the controls' routes differed slightly from DB's routes in the possible total number of turns.

For the drawing task, DB was first required to draw from memory 10 familiar objects. Subsequently, he was required to copy the same objects from a model (The models were taken from the corpus of Snodgrass and Vanderwart [1980]) to control for any drawing difficulty. DB's drawings from memory were classed as correct only if an independent observer, blind to the experiment, could name them.

Results. Table 2 shows the results for the three types of material. DB's performance did not differ from that of controls in describing familiar objects and famous faces from memory. Compared to the controls, DB's scores were slightly lower for the objects and slightly higher for the faces, but not

Table 2. Patient DB. Perceptual test scores

	DB	Controls Mean and (SD)
<i>Verbal Description</i>		
Objects	26.5	35.5 (12.6)
Faces	29.5	23.6 (7.5)
Routes (near home)	45%	98% (2.18)
<i>Drawing Objects</i>		
From memory	9/10	
Copy	10/10	
	DB	Control scores
<i>BORB Test</i>		
Objects	62/64	
Non-Objects	33/64	
Total	95/128	Mean: 115.7 Range 112–118
<i>Overlapping Figures</i>		
Meaningful	30/36	Mean: 32.4 SD: 3.6
Meaningless	6/35	Mean: 30.4 SD: 4.7

significantly so. By contrast, he showed marked difficulties in imagining routes, as shown by the low percentage of correct turns and intersections he recalled compared to near-perfect performance of the controls.

His drawings of objects from memory were all easily recognizable and his drawing skills were unaffected, as his copies were quite accurate even for relatively complex three-dimensional detail in the model objects.

Comment. Altogether, the results of verbal description tasks suggest that, except for routes, DB's internal representations of a variety of stimuli are relatively preserved. These markedly impaired perceptual skills are then probably due to disruption of processes involved in the initial evaluation of the stimuli's visual properties. This possibility is addressed below.

Object identification

We tested DB's ability to derive physical properties, such as mass, volume, and principal axis of objects under conditions of stimulus degradation. The task typically requires one to identify an object or make a decision about it. For example, identification might require recognizing objects from fragmented or overlapping line drawings or from silhouettes. A typical decision task might require deciding if a visual configuration is an object or a nonobject (see: Riddoch and Humphreys, 1987; De Renzi, 1999).

Task 2: Object decision – BORB test

Material and procedure. Riddoch and Humphrey (1987) devised this task. The material consists of 128 line drawings of which 64 are real objects and 64 are nonobjects. The nonobjects consist of a chimeric drawing consisting of two parts from different items pasted together – for example, a

horse's head pasted on the body of a squirrel. The task is simply to decide if the stimulus represents a real object or a nonobject. All stimuli are presented in free vision for the time necessary to the subject to reach a decision.

Results. DB's performance was within normal limits for real objects, but markedly impaired for the nonobjects (Table 2).

Comment. An anonymous referee suggested that the discrepancy in DB's performance between real objects and nonobjects, may have occurred as the result of a "yes" bias in DB's pattern of responding. This possibility is unlikely because, if such was the case, similar biases should have been observed also in other tasks requiring binary choices (Same-Different Faces and Buildings). This was clearly not the case in our patient. DB's performance differs from that of patient HJA, who found objects and nonobjects equally difficult (Riddoch and Humphreys, 1987). A possible explanation is that DB's relatively intact internal representations may have facilitated retention in working memory of the exemplar for the time sufficient for a successful match. A successful match could have occurred also on partial information (i.e., he might have considered only a small number of features). For the nonobjects, one would have to postulate, in addition, impairment in the verification processes. In this view, DB could have possibly terminated prematurely his verification, and thus made it less likely to detect substituted or added features in the nonobjects.

Task 3: Overlapping figure test

Material and method. The test was derived from tasks developed by Poppelreuter (1917) and Ghent (1956). In the current version (Della Sala *et al.*, 1995) two sets of line drawings were created, one set being common objects ($n = 40$) and one set meaningless shapes ($n = 36$). The stimulus item consisted of 3 to 5 overlapping drawings (targets). Below the stimulus items, there was a multiple-choice response set of nonoverlapping figures comprising the targets and the distracters. The task was to identify as many of the targets as possible.

Results. DB's performance was mildly weak on the meaningless shapes (less than one standard deviation below the mean of controls). By contrast, his score on the meaningless shapes condition was markedly impaired (over 6 standard deviations below the mean of the controls).

Comment. Again, as in the previous task DB found real objects easier. This discrepancy is also unusual (Della Sala *et al.*, 1995) and is likely again to be due to DB's relative preservation of internal representations. DB's damaged perceptual processes would engender a critical depletion of resources enabling him to generate and maintain internal representations of the meaningless shapes for the time necessary to conduct the search. On the other hand, DB's relatively intact internal representations of real objects may have made it easier for him to complete target identification even with partial information.

Taken together with the findings from the two previous tasks, results from this task suggest that most of DB's visuo-perceptual impairment can be accounted for by disruption of processes initially coding properties such as mass,

volume, and principal axis of objects under conditions of stimulus degradation.

Testing DB's complex visual stimuli processing skills

Task 4: Processing of maps, buildings and faces

The general cognitive evaluation clearly indicated that DB was able to identify countries from their outline. This was all the more surprising given his profound impairment in identifying exemplars of other complex visual stimuli (e.g., faces) and his severe visuoperceptual difficulties. As far as we know, DB did not travel extensively and it would be therefore unlikely that he had acquired specialized geographical or topographical knowledge. In the following task we first aimed to replicate and extend the finding that DB could identify maps. We used a more stringent set of stimuli than those used in the baseline evaluation. We also tested his ability to name the corresponding countries from verbal definition (as described by Incisa della Rocchetta *et al.*, 1996). Finally, we tested recognition of other complex visual materials: topographical buildings and faces. An anonymous reviewer suggested the possibility that maps are visually simpler and/or more familiar than the other stimuli. In order to control for this possibility, we adapted the Method described in Snodgrass and Vanderwart (1980) to rate visual complexity and familiarity and compared ratings for maps versus other categories of stimuli.

Material and methods. The material consisted of three sets of stimuli: 30 outline maps (of European and world countries); 32 photographs of famous buildings (again, both English and foreign); and 31 black and white photographs of famous faces (both English and foreign). Each stimulus was individually shown in free vision. All target maps were shown in the general geographic context of neighboring countries (e.g., Portugal was shown together with Spain and France). In the countries-naming task, DB was first required to name the countries from the maps and subsequently from verbal definitions (e.g., "The Stars and Stripes is the flag of what country?"). For the buildings and faces, DB was first asked to name the stimulus and, failing this, he was asked to provide as much information as possible about it. We counted as correctly identified only items on which no ambiguity was possible. Thus, "the President's house in Washington" for the White House and "former Prime Minister, Conservative Government" for Margaret Thatcher, were counted as correct but "foreign musician" was not deemed sufficient for Elvis Presley. The Method described in Snodgrass and Vanderwart (1980) to rate visual complexity and familiarity is described in the Appendix.

Results. Table 3 shows that DB has little or no difficulty in naming countries both from outline maps as well as from verbal definition. His score is only very marginally weaker than the average score of six normal control subjects of similar educational and occupational background. By contrast, his performance is very poor indeed with famous buildings and famous faces, as compared to that of four control subjects. The results of the complexity and familiarity analyses showed that, overall, maps were indeed less complex

Table 3. Patient DB. Knowledge of countries, faces, and buildings

Countries	DB	Controls Mean and (SD)		
		Named	Identified	
	DB	Controls Mean and (SD)	DB	Controls Mean and (SD)
Naming by picture	26/30	28.5 (1.4)		
Naming by definition	26/30	28.6 (1.0)		
Famous Buildings	10/32	27.0 (5.0)	18/33	28.0 (3.7)
Famous Faces	4/31	24.0 (6.3)	14/33	28.0 (5.0)

than all other stimulus types, but equally as familiar as buildings and also less familiar than famous people, animals, objects and body parts (The difference was significant for objects and body parts; see Tables A1 and A2 in the Appendix for critical values).

General comment

The aim of the four experimental tasks was to evaluate further the extent of DB's impaired visuoperceptual skills and confirm and extend the baseline finding of his intact ability to recognize maps. His apperceptive agnosia and prosopagnosia had strongly suggested that his difficulties might lie at the input stage, where structural representations of stimuli are computed. This notion is supported by his difficulties with perceptual tasks, as he clearly had problems in deriving the meaningless shapes, when these were overlapping, and in rejecting the chimeric figures.

How can he identify maps with apparent little effort, given his impairment of perceptual processes? The degree of visual complexity and richness of internal detail is unlikely to be the crucial factor in determining DB's performance in recognizing maps, as our analyses clearly show that DB had no difficulty in recognizing other visually complex stimuli, such as animals, which were rated by our control subjects as the most complex category. By contrast, DB has clear difficulties with silhouettes, which lack internal detail. Therefore it seems unlikely that his preserved ability to identify maps could be accounted for by lack of internal detail.

On the basis of these considerations, we believe that identification of maps relies not so much on shape recognition processes as on spatial analysis of the geographical location of the country in question (Incisa della Rocchetta *et al.*, 1996). In this view, coarse-grained analysis of the shape of the country might be sufficient, provided spatial processes are intact (as they indeed seem to be in DB). This hypothesis will be developed further in the General Discussion.

Case 2

Patient WH was a 66 year-old woman with a 2-year history of progressive language difficulties. She had noted increasing difficulty expressing herself and finding the appropriate name for very common objects such as birds or flowers. Her family

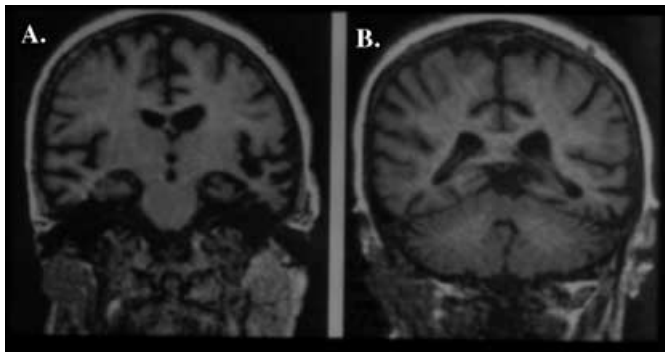


Fig. 2. MRI images of patient WH: A. Bilateral temporal lobe atrophy, more pronounced on the left temporal lobe than on the right. B. Shows normal appearance of parietal lobes bilaterally.

reported that she gradually withdrew from social contact because of difficulties in maintaining conversation. In June 1997, she was referred to Prof. Rossor at the National Hospital

for Neurology and Neurosurgery for further investigation of her symptoms. Neurological examination was normal except for her cognitive deficits, which are described in detail below. Structural neuroimaging (MRI) showed marked bilateral focal anterior temporal lobe atrophy, greater on the left (Fig. 2A) with relative sparing of the posterior temporal lobes bilaterally and normal parietal lobes (Fig. 2B).

Neuropsychological assessment

WH underwent two formal neuropsychological assessments (July 1997; April 1998); the results are shown in Table 4.

First assessment (July 1997). At the time of her first assessment, she obtained a defective score on the Verbal scale of the WAIS-R and an average score in the Performance scale, indicating severe intellectual deterioration on the Verbal scale. Her verbal short-term memory functions were relatively preserved as she obtained a digit span of five digits forward.

Table 4. Patient WH. Baseline cognitive scores

	1st assessment 1997		2nd assessment 1998	
<i>General Intelligence</i>				
VIQ	68		not tested	
PIQ	92		not tested	
<i>Memory</i>				
<i>Recognition Memory Tests</i>				
Words	19/25	at 5 th %ile	not tested	
Faces	34/50	<5 th %ile	not tested	
Easy Faces	not tested		13/25	chance
Easy Buildings	not tested		22/25	at 5 th %ile
<i>Famous Faces</i>				
Named	0/12		not tested	
Recognized	1/12	<5 th %ile	not tested	
<i>Language</i>				
<i>Naming</i>				
Nouns (GNT)	0/30		not tested	
Nouns (Patterson <i>et al.</i> , 1994)	not tested		1/106	
Verbs	not tested		5/30	
<i>Comprehension</i>				
<i>Visual-visual Matching</i>				
Animals	not tested		23/50	
Objects	not tested		40/50	
<i>Pyramids and Palm Trees Test</i>				
Pictures	not tested		32/52	<46 cut off
Words	not tested		30/52	<46 cut off
<i>Reading, Spelling, Calculation</i>				
<i>Reading</i>				
NART	0/50		not tested	
Schonell	41/100	Reading IQ < 78	not tested	
Regular	not tested		43/53	
Exception	not tested		14/53	
non-words	not tested		17/20	
<i>Spelling</i>				
GST	2/30	<5 th %ile	not tested	
<i>Calculation</i>				
GCT	4/24	<5 th %ile	not tested	
<i>Perception</i>				
Incomplete Letters	19/20	Above 5% cutoff	not tested	
Cube Analysis	10/10		not tested	

GNT = Graded-difficulty Naming Test.
 GST = Graded-difficulty oral Spelling Test.
 GCT = Graded-difficulty oral Calculation Test.
 %ile = percentile.

Forced-choice recognition memory (Warrington, 1984, 1996) was impaired for both words and faces. She was unable to name and recognize famous faces (Warrington and James, 1967).

Her language functions were profoundly impaired. Her spontaneous speech was fluent, well articulated, but very poor in informational content, as it was limited to a few stereotyped sentences. On formal testing, she presented with a grave nominal dysphasia, as she was unable to name any item on a stringent graded difficulty object-naming test (McKenna and Warrington, 1983). Her comprehension was also severely affected. This aspect was explored in more detail at the time of her second assessment.

Her reading of single words was severely impaired. She was unable to score on the National Adult Reading Test (NART), and she scored poorly on the Schonell Reading Test (Schonell, 1942). Several regularization errors were noted, suggesting surface dyslexia. This is often reported in patients with semantic dementia (but see Cipolotti and Warrington, 1995). Her reading of single letters was preserved.

WH's spelling skills were also severely impaired, as she was barely able to score on the Graded Difficulty Oral Spelling Test (Baxter and Warrington, 1994). In a writing-to-dictation task, using regular and exception words (Patterson and Hodges, 1992), her performance was equally poor for both the regular (59%) and the exception words (47%). Her calculation skills were also affected, as demonstrated by her poor performance on a Graded Difficulty Oral Calculation Test (Jackson and Warrington, 1986).

By contrast, her visuo-perceptual and visuo-spatial skills were well preserved, as is clearly indicated by her nearly flawless performance on the Incomplete Letters Test of visual perception and Cube Analysis Test of visuo-spatial perception (Warrington and James, 1991).

Second assessment. In April 1998 WH presented with further deterioration of her cognitive skills. In the second neuropsychological assessment, we focused on her visual memory and language functions (Table 4). We administered two relatively easy recognition memory tasks, one for unfamiliar faces, the other for unfamiliar buildings (Clegg and Warrington, 1994). These tests are roughly matched for difficulty. WH's performance was at chance on the faces, but relatively creditable on the buildings. This raises the possibility of some preservation of topographical memory.

WH's language skills had further deteriorated with respect to the first assessment. Her naming was virtually abolished, irrespective of whether nouns or verbs were used. Her comprehension skills were assessed on a visual-visual matching task (Cipolotti and Warrington, 1995) and on the Pyramids and Palm Trees Test (Howard and Patterson, 1992). WH was profoundly impaired in comprehending both pictures and spoken words. In the visual-visual matching task, her performance was very poor for both animal and object categories. In the Pyramids and Palm Trees Test, she performed poorly on both pictures and words.

Her performance was also gravely impaired on a test in which she was required to give verbal definitions of 20

animals (elephant, gorilla) and 20 objects (helicopter, television). The same items were presented both in the verbal or visual modality. In the verbal modality, the names of the objects were printed on cards. In the visual modality, she was shown realistic toy objects. WH was virtually unable to produce any recognisable definition irrespective of the modality of presentation of the items (Verbal modality 0/40; visual modality 4/40). Qualitatively her errors in the verbal definitions suggested associative agnosia. For example, she said about a brush "where you push it in water." She was also requested to point on verbal command to each one of the toys in turn (within arrays of 5). She was barely able to score on this task (5/20).

WH's reading skills were reassessed to ascertain if she was differentially sensitive to the regularity or frequency of words. We employed a test designed by Graham *et al.* (1994). The stimuli consisted of 106 imageable words, of which 53 were regular and 53 were exception words matched for frequency and syllable length. The regular-exception pairs were further classified as belonging to three frequency bands: High ($n = 28$), medium ($n = 36$) and low ($n = 42$). For further details see Graham *et al.* (1994). WH showed a clear regularity effect, but no frequency effect as her correct responses were roughly equally distributed across the frequency bands (High = 16/28; Medium = 19/36; Low = 22/42); WH's ability to read monosyllabic and disyllabic nonwords was relatively well preserved.

Comment. WH's language difficulties were characterized by progressive fluent aphasia, with a severe impairment of naming and comprehension. Surface dyslexia and surface dysgraphia were also present while her visual perceptual and visuo-spatial skills were preserved. She presented with bilateral temporal-lobe atrophy, more marked on the left side. The provisional diagnosis of semantic dementia was reached for WH, as her symptoms clearly fit the pattern observed in typical cases with respect to the neurological, neuropsychological and neuroradiological profiles (Snowden *et al.*, 1989; Hodges *et al.*, 1992, 2000; Mummery *et al.*, 2000; Chan *et al.*, 2001). Semantic dementia is a subtype of frontotemporal lobar degeneration (Neary *et al.*, 1998).

Experimental investigations

Given WH's profound comprehension impairment, we explored whether she presented with selective preservation of some categories of knowledge. This was motivated by recent reports documenting that patients with semantic dementia may show selective sparing of some semantic categories. For example, selective sparing of the categories of color (Robinson and Cipolotti, 2001) number (Cappelletti *et al.*, 2001) or body parts (Shelton *et al.*, 1998) has been recently demonstrated in patients with semantic dementia. Preservation of map knowledge has been also reported in at least four cases with semantic dementia. Interestingly, preservation of map knowledge was associated with preservation of topographical memory in at least one case (patient BF; Maguire and Cipolotti, 1998; Cipolotti, 2000). In this context,

it should be noted that our patient showed a somewhat better memory for unknown buildings than for unknown faces. This finding hints at the possibility that she may show some preservation of topographical memory. In view of the previous reports we decided to investigate comprehension of a variety of categories of knowledge. In particular we have explored in further detail her comprehension of topographical material. As was the case for DB, we should stress that, as far as we know, WH did not travel extensively and it would be therefore unlikely that she had acquired specialized geographical or topographical knowledge. All testing was carried out during WH's second assessment, carried out in April 1998.

Task 1: Naming, transcoding and comprehension for different semantic categories

The purpose of the present task was to assess the extent of WH's naming, transcoding (i.e., reading, writing, and repetition), and comprehension skills for different semantic categories.

Material and method. There were five categories of nouns: 10 well-known countries, 10 colors, 10 animals, 10 objects, and 10 body parts (partly based on McKenna and Warrington, 1978; see Cipolotti, 2000 for further detail). The stimulus names were matched for frequency (A and AA; Thorndyke-Lorge, 1944), and length, except for country names, which were slightly longer (countries mean length = 6.2, SD = 1.1; animals = 4.3, SD = 1.15; objects = 4.3, SD = 0.8; body parts = 4.0, SD = 0.9; colors = 4.7, SD = 0.9). There were eight conditions of testing: (1) oral and (2) written naming by confrontation, (3) reading aloud, (4) writing to dictation, (5) repetition, (6) spoken word-picture matching same category (blocked), (7) spoken word-picture matching in random order, and (8) written word-picture matching same category (blocked). In the naming tasks, WH was asked to say or to write the name corresponding to a picture (naming by confrontation). In the reading task, WH was asked to read single words aloud, each typed on a card. In the writing to dictation task, the examiner said each word once and WH was asked to write it down. In the word repetition task, the examiner said

each word once and WH was asked to repeat the stimulus immediately. The same stimuli were used in all sessions and were always presented in blocks of five items, one from each category. The stimulus set was very easy for control participants, as shown a previous investigation. The same stimuli used here were administered in a written naming condition to five control participants. Their performance was at ceiling (10/10) in all categories (Cipolotti, 2000).

Her comprehension skills were tested using matching-to-sample techniques in which WH was required to match either a word (spoken or written) to a stimulus item in an array displayed on a desk. Each stimulus was probed once in 10-item arrays. In the random condition the arrays comprised items randomly selected from the five categories whereas in the blocked condition, the 10 items were from a single category. All items were tested twice, over four separate sessions on two consecutive days in the following order: Day 1 – Session I: writing to dictation (Trial 1), spoken word-picture matching in random order, oral naming by confrontation. Day 1 – Session II: reading aloud, written naming by confrontation. Day 2 – Session I: written word-picture matching same category (blocked), spoken word-picture matching same category (blocked). Day 2 – Session II: repetition, writing to dictation (Trial 2). This task has been validated in previous work and it proves very easy for control subjects, who perform at ceiling in virtually all categories and all conditions (Cipolotti, 2000).

Results. Table 5 shows that WH's performance in the two naming tasks was practically at floor for all categories except for countries and colors, in which she performed only marginally better. The pattern of her errors in both naming conditions was very consistent. Thus, most of her errors were omissions (67%) followed by semantic errors (21%). Very few instances of other types of errors were noted (3% phonological errors, e.g., "north" for nose and "sparr" for Spain; 4% perseverations; 10% circumlocutions; 4% unrelated). Her ability to read aloud was relatively spared in contrast to her performance on writing to dictation, which was impaired. Repetition was virtually flawless (96%).

Table 5. Patient WH. Naming, transcoding, and comprehension for different semantic categories

	Countries	Colors	Animals	Objects	Body parts	Total (/100)
Naming, N. correct (/20)						
Oral Naming	5	9	2	0	3	19
Written Naming	7	5	2	0	1	15
Percent	30	35	10	0	10	17
Transcoding, N. correct (/20)						
Reading aloud	19	20	15	19	16	89
Writing to dictation	11	13	10	9	5	48
Repetition	19	20	19	20	18	96
Percent	81.6	88.3	73.3	80	65	77.6
Comprehension, N. correct (/20)						
Spoken-word/picture matching Blocked	18	8	6	7	6	45
Spoken-word/picture matching Random	16	18	12	9	12	67
Written-word/picture matching Blocked	18	15	7	9	12	61
Percent	86.6	68.3	41.6	41.6	50	57.6

In the spoken and written word-picture matching conditions her performance was gravely impaired across all semantic categories, with one remarkable exception: the category of countries. Thus, she was able to match the names of countries with the corresponding map in more than 85% of her attempts. By contrast, her performance was at or below 50% for most other categories (except colors: 68%).

Comment. The results obtained delineate the extent of WH's semantic memory impairment. Her naming skills were practically abolished. Her comprehension impairment was severe and affected almost all categories of knowledge tested. Living things, objects, and body parts were indeed all equally and gravely affected. The only remarkable exception was the relative sparing of the category of countries. Her comprehension of countries was well preserved in all comprehension conditions, irrespective of modality of stimulus presentation (spoken or written) or order of presentation (random or blocked). The pattern of preservation of countries replicates and extends previously reported findings obtained with identical materials and procedures (Cipolotti, 2000). The fact that countries were selectively impaired in another patient (patient DK, Incisa della Rocchetta *et al.*, 1996) again, with identical materials and procedures makes it unlikely that this category is easier than the others in the same set.

WH is the first documented case in whom selective sparing of the categories of countries is clearly demonstrated only in the comprehension condition. In all previously published cases, sparing was observed in both naming and comprehension (e.g., patient TM: Incisa della Rocchetta *et al.*, 1998; patient BF: Cipolotti, 2000; patient SA: Ward *et al.*, 2000).

Task 2: City-country association

The next step was to test further categories of visual knowledge in light of the dissociations observed in previously published cases between knowledge of countries and other types of visual information. For example, patient APA (Miceli *et al.*, 2000) had preserved map comprehension, but had some loss of knowledge of famous people. By contrast, patient DK, was profoundly impaired in recognising countries, but had spared visual knowledge of famous buildings, historical people, and contemporary celebrities (Incisa della Rocchetta *et al.*, 1998). With these findings in mind, we tested WH's associative knowledge of cities (i.e., when paired with names or maps of countries).

Material and method. There were two conditions: verbal/verbal and verbal/visual. In the verbal/verbal condition, the written names of 12 cities were each paired with the names of two alternative countries, one being the country in which the city was located (e.g., is London in England or in Ireland?); the patient was requested to select the correct alternative. Each triad was probed twice. In the verbal/visual condition the procedure was identical, except that the two alternative countries were presented in map form. It can be clearly seen (Table 6) that her performance was rather poor in both conditions, being barely above chance. By contrast, four control participants, matched in age gender and educational

Table 6. Patient WH. Associative knowledge of cities, buildings, and familiar faces with countries

	Type of association			
	Verbal-verbal		Verbal-visual	
	WH	Controls	WH	Controls
City-country	15/24	12/12	15/24	12/12
	Verbal-verbal		Visual-visual	
	WH	Controls	WH	Controls
Building-country	14/20	10/10	19/20	10/10
Famous face-country	11/20	10/10	9/20	10/10

level to WH, performed flawlessly (items were presented only once to the controls).

Task 3: Location of cities

In this task we probed knowledge for location of cities within an outline map.

Material and method. WH was required to indicate on a blank map of Britain the position of 15 amongst the major cities. The cities' names were either: (1) spoken by the examiner or: (2) printed on cards.

Results. She was basically unable to score on either version of this task, obtaining 2/15 on both conditions. Eight of her errors were omissions. The four remaining errors were all quite gross, with Belfast, York, Newcastle and Manchester, all being placed in a cluster in Southeast England.

Comment. WH's poor performance on the last two tasks documents her severe loss of knowledge for cities – both associative and spatial knowledge were virtually abolished. Thus she could no longer associate a city with the corresponding country. The pattern of errors in the location task indicates that she did not have even a coarse-grained knowledge of position of the cities (e.g., north versus south).

Task 4: Building-country association

Material and method. In this task, knowledge of world-famous buildings was probed both verbally and visually. In the verbal condition, the written name of a famous building (e.g., Statue of Liberty) was presented together with the written names of two countries, one of which is the country where the building is located (e.g., USA, Canada). WH was requested to point to the correct alternative, each stimulus set was probed twice (once in the controls). The visual condition was identical in procedure except the stimuli were photographs of the same famous buildings and outline maps of the countries.

Results and comment. WH's performance (Table 6) was impaired in the verbal condition and almost flawless in the visual one. It is possible that her visual knowledge for famous buildings might be relatively preserved, although the fact that

the controls scored at ceiling does not allow one to conclude this with certainty.

Task 5: Famous face-country association

This task's aim was to ascertain whether WH's residual semantic knowledge extended to famous people.

Material and method. Knowledge of famous people was probed both verbally and visually. In the verbal condition, the written name of a famous person (e.g., Gandhi) was presented with the written names of two countries, one of which was the country associated with the famous person (e.g., India, Germany). In the visual condition a photograph of the same famous person was paired with the maps of two countries. In both tasks, WH was required to point to the country of origin of the famous person.

Results and comment. WH performed at chance on both the verbal and visual modalities whereas the control participants all performed flawlessly (Table 6). This suggests that knowledge of famous people was impaired. These results contrast with those obtained in the previous task, which showed relative preservation of visual building knowledge. This dissociation supports the notion that the systems in the brain coding faces are separate and independent from those coding other types of visual information (see De Renzi, 1999; Desimone, 1991 for discussion of findings in patients and monkeys, respectively), including topographical and geographical knowledge (Incisa della Rocchetta *et al.*, 1996).

Task 6: Face familiarity

This task aimed to explore if WH's face processing impairment was at the level of associative links between the faces and the countries or at the level of the representation of the faces themselves.

Material and method. In this task, 15 faces of famous people were presented individually, each paired with two distracter faces. These were faces of unfamiliar people, selected to resemble closely the target face. WH was required simply to discriminate the familiar face from the unfamiliar ones (Warrington and McCarthy, 1988).

Results and comment. WH's performance was at chance on this test (6/15). This indicates that her poor performance on the previous task is underpinned by a basic deficit at the level of the representations of the faces.

General comment

Task 1 has demonstrated that WH has a profound loss of knowledge for a variety of categories of common names. The only exception was her preserved knowledge of countries, which was the only category spared within the more general category of proper nouns. WH presented also with a relative preservation of famous buildings. This contrasts with her impairment for two other categories of proper nouns: famous people and cities. These findings are one further illustration of the variety of ways in which knowledge can fractionate. They support the notion that the representation of countries appears

to be separate and independent from the representations of faces.

Discussion

The present study focused on the preservation of a category of knowledge: countries. We presented the findings of two patients. The aetiology was vascular in the first patient, degenerative in the second. In the first patient, DB, the ability to recognize and name countries from their map outline was preserved, despite severe visual perceptual deficits. In the second patient, WH, comprehension of map outlines indicated greater residual knowledge of the category of countries in the context of pervasive disintegration of semantic knowledge.

Detailed cognitive testing showed that patient DB presented with dense apperceptive agnosia and prosopagnosia. He was very sensitive to various types of visual transformations of stimuli. His performance was significantly impaired when contours were partially obliterated (Incomplete Letters), texture and volume information reduced (Object Decision, Silhouettes) and typical orientation of the objects changed (Unusual Views). He had marked difficulties in rejecting chimerical nonanimals (BORB test; e.g., the body of a dog with the head of a cow) and he was severely impaired in matching a nonsense shape to its replica (Overlapping Figures). By contrast, his performance in identifying objects was preserved when they were presented as perceptually nondegraded images in a canonical perspective. In addition, he could draw recognizable exemplars of objects from memory and provide reasonable descriptions of objects and faces.

Famous faces and buildings, on the other hand, proved very difficult to identify for DB, even when the images were not degraded in any way. He had severe difficulty even in *matching* a photograph of a face or of a building, with the same face or building photographed from a different (still canonical) perspective. The remarkable finding in this patient is that, despite such severe and pervasive visual perceptual deficits, he had no difficulty whatsoever in identifying countries from their outline maps. A simple explanation of this category effect in terms of visual complexity appears unlikely. Maps were found to be less complex than other classes of visual stimuli. However, we saw that he had no difficulty visually recognizing items that were more complex than maps (e.g., animals, objects, body parts).

How can we explain this preservation? How do the present findings bear on the three major theoretical positions discussed in the introduction?

We are going to argue that the findings obtained in patient DB are problematic for the account put forward by Humphreys and collaborators (Riddoch and Humphreys, 1987; Humphreys *et al.*, 1988; Forde *et al.*, 1997). This view postulates that category-specific effects (especially the category-specific impairment for living things) originate at a presemantic stage, namely at the stage in which structural description is accessed. At this stage, information derived from perceptual processes is mapped on stored information

(internal representations) in the structural description stage. In the structural description stage, information is vulnerable to errors. According to the authors, this is likely to occur especially when a perceptual category contains many similar, competing, exemplars. This is due to a spread of activation across the structural representation of perceptually similar neighbors (Humphreys *et al.*, 1988, 1997; see also Caramazza *et al.*, 1990; Gaffan and Heywood, 1993; Rapp *et al.*, 1993; Hillis *et al.*, 1995).

How does interference at the level of structural description affect later stages? The central assumption of this account is that object processing can propagate forward to several stages, in *cascade* fashion, before processing is complete in the earlier stages. This increases the likelihood that the representations of competing stimuli reach the semantic or phonological stages before the representation of the target stimulus (see Gale *et al.*, 2001). Consequently, a category-specific effect appearing to originate at the semantic stage could have occurred as the result of interference at the level of structural description.

It is generally accepted that object recognition processes heavily rely on shape and texture information. Hence, spread of activation at the structural description stage should mainly affect shape and texture information. Therefore, categories with similar shapes and textures, such as, for example animals (e.g., trunk shape with head, legs, tails, and fur texture), faces (e.g., oval shape with eyes and nose, skin texture), or buildings (e.g., parallelepipedal shape, with doors and windows, brick, stucco, or cement texture) should be particularly vulnerable to interference effects at the structural description stage. This prediction is not upheld in DB. Although he has difficulties in recognizing faces and buildings and performs poorly on tasks requiring processing of shape and texture, he clearly can perceive normally some categories relying on shape and texture information, such as animals and body parts. The items in these categories were also rich in internal detail. That richness of internal detail cannot be a crucial factor is also supported by DB's good performance in recognizing objects in the control condition of the Unusual Views test. These objects are the very same ones that prove difficult for DB to identify in the unusual perspective. Yet the photographs do not differ in any respect other than perspective. Crucially, the photographs are grainy and rich in shadows and internal detail.

On the basis of these considerations, we think that our findings somewhat undermine the theoretical assumptions of the account of Humphreys and collaborators. Although this model saw its genesis in trying to explain the specific impairment of living things, its general assumptions may be generalized to other categories comprising visually similar exemplars (e.g., faces, buildings, and maps). The model's predictions that perceptual deficits involving the level of structural descriptions should affect categories relying on shape and texture information is clearly not upheld here as countries, animals, and body parts are preserved in patient DB. Moreover, the findings in our second patient, WH also appear problematic for this account.

Patient WH, who suffered from semantic dementia, showed a severe impairment in naming and comprehension of both living and nonliving things, such as animals, objects, as well as of other categories. By contrast, comprehension (and naming to a lesser extent) of countries was relatively spared. Detailed testing unveiled a pattern of associations and disassociations between the category of countries and other categories of proper nouns. Thus, WH had also partially spared knowledge of buildings in the visual modality. By contrast, knowledge of cities and faces was consistently impaired, regardless of modality. WH is the first patient on record, to our knowledge, to demonstrate specific sparing of the category of countries at the level of comprehension; all patients reported to date show preservation at the level of naming. It is likely that this is due to her impairment being more severe than that observed in the other published cases. These patients generally showed preserved comprehension skills across several semantic categories (see Cipolotti, 2000).

What are the implications of the findings in patient WH for current accounts of category-specific impairment? In our opinion, they are problematic for the view that category-specific effects occur outside the semantic system, at a pre-semantic perceptual stage (Riddoch and Humphreys, 1987; Humphreys *et al.*, 1988; Forde *et al.*, 1997). WH showed a profound impairment for certain visually complex and *crowded* categories, such as animals and faces, whereas other visually complex and crowded categories, such as buildings, were spared, although buildings were significantly more complex than countries. This is clearly in contradiction with this account, as it predicts roughly equivalent levels of impairment for all visually complex and crowded categories. One may argue that countries also need subtle distinctions in shape to distinguish one from the other – think of Africa, India, and South America with their roughly triangular shape, although countries may have fewer competing exemplars than other categories (e.g., buildings, animals, and faces).

In view of the unlikelihood of a perceptual account, we postulate that the effects we observed in WH originate from damage in the semantic system. As discussed in the introduction, two major hypotheses regarding the organization of categories of knowledge within the semantic system have been put forward. The position of McCarthy and Warrington (1990) holds that categories are acquired during ontogenetic development. The position of Caramazza and collaborators holds that categories are acquired during phylogenetic development (Caramazza and Shelton, 1998; Caramazza, 2000).

According to McCarthy and Warrington, the way in which categories are acquired in childhood would determine how they are encoded and stored in the brain. In particular, this account hypothesized a motor channel as underpinning the development of categories of artefacts (as the child acquaints himself or herself with artefacts through use and manipulations) and sensory channels as underpinning the development of animal categories (distinguishing between animal categories engenders subtle distinctions of shape and texture). This framework is too broad, however, to account for idiosyncratic

effects such as selective impairment or sparing of map knowledge. We extended its basic assumptions to accommodate the effects of the selective impairment and preservation of maps (Incisa della Rocchetta *et al.*, 1998; Cipolotti, 2000). We proposed that when first learned, maps do not engage visuo-motor channels like objects that one manipulates, or objects that one walks around, like buildings. Whereas manipulable objects, faces, and buildings have a definite and estimable size, maps of countries are unfathomable in size. They are learned from atlas representations in variable, arbitrary scales. Moreover, maps occupy fixed spatial configurations, a property shared by buildings but not by most objects and not by faces, which are seen in a variety of different contexts. We hypothesized that maps are represented in a coarse-grained spatial pattern which respects topological relations but does not retain precise metric information. It is this spatial context (i.e., neighboring countries) which we believe is crucial for identification of the country. Our findings lend support to this hypothesis as both our patients had intact spatial processes. Of particular interest for our spatial hypothesis are the findings in patient DB. This patient had profound deficits in shape perception; however, he had no difficulty in perceiving spatial location. Thus, it seems possible that he made use of the spatial context to boost recognition of maps. Hence, we speculate that had we presented only the contour of the target country (say, Portugal, without neighboring Spain and France), DB would have had serious difficulties identifying the countries. Unfortunately we did not carry out this task and therefore we cannot exclude that some countries could have been recognized by shape alone. However, we believe that buildings, like countries, but unlike most other categories of objects, are also part of a spatial configuration with fixed relationships with neighboring buildings. We think it is this spatial context that may boost recognition of the individual building. For example, the *Arc de Triomphe* is situated in the round-shaped *Place de l'Etoile* with radiating avenues and trapezoid-shaped buildings. This context might help one identify the Arc in a photograph and distinguish it more easily from similar triumphal arches in other European capitals. Indeed, we believe that these topological and topographical cognitive processes ultimately engage brain regions different from those engaged by objects and faces and this is consistent with anatomical findings in patients with selective sparing of the category of countries.

To our knowledge, except for one patient (patient AD, probable Alzheimer's Disease; Incisa della Rocchetta *et al.*, 1998) all patients showing selective sparing of the category of countries suffer from semantic dementia. This condition affects the temporal lobes bilaterally, but with greater left-sided damage and with an anterior-posterior gradient, with more marked damage anteriorly (e.g., Mummery *et al.*, 2000; Chan *et al.*, 2001). Parietooccipital regions in the nondominant hemisphere are typically spared. This was also the case in patient WH. Indeed, in previously reported patients showing selective sparing of the categories of countries, the posterior region of the right hemisphere was relatively preserved (see

Cipolotti, 2000). This has led to the suggestion that the sparing of countries could be explained as a consequence of sparing of the specialised neural systems that represent and compute spatially based topological representations (Incisa della Rocchetta *et al.*, 1998). A speculation was offered that these spatially based topographical representations may be closely linked to other spatial skills represented in the posterior part of the right hemisphere (Cipolotti, 2000), suggesting that these areas play a critical role in map knowledge. In line with this, one of us (LC) reported in a recent functional neuroimaging study that processing maps recruits the posterior regions of the nondominant hemisphere, specifically the right occipitoparietal and parahippocampal cortices (in addition to the left extrastriate cortex; Gorno-Tempini *et al.*, 2000). The localization of the activations was interpreted to be reflecting the spatial nature of topological representations. In this study dissociations between the regions activated by maps, buildings, and faces were also reported.

The anatomical findings in DB allow further development of this account. This patient presented with a right parietal and bilateral occipital infarcts (right larger than left). His lesions, however, appear largely to spare the more dorsal aspect of the parietooccipital cortex, which is believed to be particularly involved with spatial processes (Burgess *et al.*, 1999). We postulate that the critical region for maps is represented within this more dorsal aspect of the parieto-occipital cortex (cf. Fig. 2 with Gorno-Tempini *et al.*, p. 1256, Fig. 2b) and might include the parahippocampal cortex as well. Interestingly, the latter receives connections from the posterior parietal region (Seltzer and Pandya, 1984). In this light, it should be noted that in patient DB there is no discernible damage in the right medial temporal region and that in patient WH the atrophic process seems to have largely spared this region or, at the very least, only affected it minimally.

The hypothesis that different categories can be mapped onto separate brain regions has recently received strong criticism by Devlin *et al.* (2001). The authors' literature review on category-specific effects, in both functional imaging and brain lesion studies, indicated that the available evidence was rather inconsistent and did not allow one to conclude that a given category of knowledge would recruit neuronal space different from that engaged by other categories. Whilst we do not deny that Devlin *et al.*'s criticism may apply to many of the findings in the literature, our claim is that the category of countries represents a special case. Both the structural imaging data in patients and functional imaging findings in normal subjects seem to suggest that this is the case.

Turning now to discuss the position of Caramazza and collaborators (Caramazza and Shelton, 1998; Caramazza, 2000), the authors suggest that separate brain mechanisms have evolved to process categories of knowledge that are crucial for survival. These processes would allow one to distinguish food-stuff from harmful substances, friend from foe and possibly tools from one another. However, Caramazza and Shelton's view does not address how the brain might deal with categories of knowledge dictated by cultural, rather than biological,

evolution. Countries are a typical example. Countries and maps have appeared very late in the history of biological evolution and it is unlikely that a prewired, dedicated brain module has had the time to evolve during phylogenesis. Thus, in this context, the formulation by Caramazza and colleagues would have difficulty explaining the *countries* effect.

Category-specific effects remain one of the most puzzling findings in neuroscience, especially because they are so difficult to fit into a biologically plausible framework. Caramazza and Shelton's approach goes a long way in that direction but stops short of the goal in light of the difficulties in explaining some of the most peculiar effects, including those documented here. We believe a possible way forward is offered by the concept of *neoteny*, which holds that evolutionary progress in human beings has occurred through the retention in adulthood of some of the juvenile characteristics of preceding species (Gould, 1977, 1981; Montagu, 1989; see also Bjorklund, 1997). With regards to brain development, neoteny would cause retardation of brain maturity in ontogenesis. This, in turn, would allow for greater plasticity to changing environmental demands and for the development of categories of knowledge suited to a changing environment (Mithen, 1996). Support for this hypothesis is provided by simulation experiments, showing that categories of knowledge can form plastically in a self-organizing neural network, using unsupervised learning (Ritter and Kohonen, 1989). Interestingly, the resulting semantic neural space self-organizes into hierarchically nested domains, which are topographically separate and identifiable within the network. The topographical arrangement mirrors the diversity and separateness of the concepts.

We hypothesize that it is precisely this indeterminacy and plasticity which may allow for the development of idiosyncratic categories such as countries. This does not preclude that categories that have maintained crucial survival value throughout the evolution of the human species may have indeed formed during phylogenesis and may have acquired dedicated brain modules. We propose, however, that there is a portion of neural space which is available to accommodate any new categories that gain relevance during ontogenetic development.

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Appendix

Complexity and familiarity ratings

We tested 10 subjects amongst work colleagues and friends of one of us (AIdR) with an average of 42.4 years of age and 15 years of education. Complexity was defined as depending on the amount of detail and the intricacy of line in the *whole picture*, including the background (counting the lines, shadows and/or colors). Familiarity was defined according to how usual or unusual the picture in the person's realm of experience was and to which degree the person came in contact or thought about the item represented in the picture. Each stimulus was rated on a five-point scale. For complexity, 1 was very simple

and 5 very complex. Likewise, for familiarity, 1 was very unfamiliar and 5 very familiar. We analyzed ratings for maps, famous faces, and buildings, as well as objects, animals, and body parts from the naming task mentioned in DB's baseline Neuropsychological Assessment (on which he had no difficulty). One-way ANOVAs, with stimulus type as the grouping factor and subjects' ratings as the dependent variable, showed significant effects of stimulus type (Complexity: $F(5, 117) = 95.76$, $p < 0.001$; Familiarity: $F(5, 117) = 15.1$, $p < 0.001$). Post hoc analyses showed significant effects (Tukey HSD, with significance set at $p < 0.01$, to minimize the risks of Type I error). Tables A1 and A2 show the comparisons for Complexity and Familiarity ratings, respectively.

Table A1. Complexity ratings: Tukey HSD comparisons at $p < .01$

Category	Versus	Observed difference	Critical difference	
Maps	Objects	.843	.541	S
	Body	1.463	.541	S
	Famous People	1.588	.380	S
	Buildings	2.112	.377	S
	Animals	2.393	.541	S
Objects	Body	.620	.663	
	Famous People	.745	.539	S
	Buildings	1.269	.537	S
	Animals	1.550	.663	S
Body	Famous People	.125	.539	
	Buildings	.649	.537	S
	Animals	.930	.663	S
Famous Faces	Buildings	.524	.374	S
	Animals	.805	.539	S
Buildings	Animals	.281	.537	

S = Significantly different at $p < .01$.

Table A2. Familiarity ratings: Tukey HSD comparisons at $p < .01$

Category	Versus	Observed difference	Critical difference	
Buildings	Maps	.013	.567	
	Famous People	.462	.553	
	Animals	.813	.801	S
	Objects	1.193	.801	S
	Body	1.563	.801	S
Maps	Famous People	.449	.567	
	Animals	.800	.811	
	Objects	1.180	.811	S
	Body	1.550	.811	S
Famous Faces	Animals	.351	.811	
	Objects	.731	.801	
	Body	1.101	.801	S
Animals	Objects	.380	.801	
	Body	.750	.989	
Objects	Body	.370	.989	

S = Significantly different at $p < .01$.