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Knowledge of famous faces and names in semantic dementia

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Summary

Semantic dementia is a focal clinical syndrome, resulting from degeneration of the temporal lobes and characterized by progressive loss of conceptual knowledge about the world. Because of the highly circumscribed nature of the disorder it is a natural model for improving understanding of how semantic information is cerebrally represented. There is currently a lack of consensus. One view proposes the existence of modality specific meaning systems, in which visual and verbal information are stored separately. An opposing view assumes that information is represented by a unitary, amodal semantic system. The present study explores these alternatives in an examination of famous face and name knowledge in 15 patients with semantic dementia. The study of face recognition in patients with an established semantic disorder also permits an examination of the relationship between semantic dementia and the focal clinical syndrome of progressive prosopagnosia. The semantic dementia patients were profoundly impaired on both face and name identification and familiarity judgement tasks compared with amnesic patients with Alzheimer's disease and healthy controls. However, whereas the two reference groups performed better for names than faces, the semantic group showed the opposite pattern. This overall profile masked individual differences: semantic dementia patients with predominant left temporal lobe atrophy showed better recognition of names than faces, whereas patients with right temporal predominance showed the reverse pattern. Relative superiority for names or faces was mirrored by corresponding superiority for words or pictures on a standard semantic test. We interpret the findings as inconsistent with a unitary, amodal model of semantic memory. However, the data are not wholly compatible with a strict multiple system account. The data favour a model of semantic memory comprising a single interconnected network, with dedicated brain regions representing modality specific information. The data emphasize the importance of the anterior, inferolateral parts of the left temporal lobe for the representation of names and the corresponding parts of the right temporal lobe for faces. Dissociations between face and name knowledge provide a challenge for existing models of face processing. Moreover, they lead us to argue that the focal syndrome of progressive prosopagnosia is one of the clinical presentations of semantic dementia and not a separate clinical entity.

Keywords: semantic dementia; Alzheimer's disease; face recognition; name recognition; prosopagnosia

Abbreviations: FRUs = face recognition units; MMSE = Mini Mental State Examination; PINs = person identity nodes; VOSP = Visual Object and Space Perception Battery

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Introduction

Focal forms of cerebral degeneration may give rise to remarkably circumscribed patterns of neuropsychological deficit. Perhaps the most striking is the selective disorder of semantic memory that occurs in association with focal degeneration of the temporal lobes, referred to as semantic dementia (Snowden *et al*., 1989; Hodges *et al*., 1992). The disorder is characterized by profound anomia, with semantic errors (e.g. 'dog' for rabbit; 'water' for milk) and impaired word comprehension (e.g. Doctor: 'You can put on your jacket'; Patient: 'jacket, what's jacket?'). However, the semantic deficit is not confined to the verbal domain. Patients may exhibit difficulties in recognizing faces, objects, smells, tastes and non-verbal environmental sounds, such as the ringing of a telephone or rainfall on the window-pane

(Snowden *et al*., 1996; Bozeat *et al*., 2000). The term semantic dementia was introduced (Snowden *et al*., 1989) to encapsulate the multi-modal nature of the semantic disorder. Neuroimaging (Mummery *et al*., 2000; Chan *et al*., 2001) and pathological studies (Snowden *et al*., 1996) show temporal lobe atrophy, which is bilateral although often with greater emphasis on the left or right. The inferior and middle temporal gyri are predominantly affected. The hippocampi are relatively preserved, accounting for the preservation of day-to-day memorizing that is a striking feature of the disorder. Histologically brains typically show neuronal loss, severe microvacuolation with mild astrocytic gliosis, consistent with the microvacuolar form of histology described in frontotemporal dementia (Lund and Manchester groups, 1994; Mann *et al*., 1993). Histological features of Alzheimer's disease are absent. Semantic dementia is theoretically important because it provides a relatively pure model for exploring the psychological organization and neural representation of semantic memory.

Multiple semantics or an amodal semantic system?

It is well recognized that, although semantic loss may encompass different modalities of information, it does not invariably do so equally. Patients with impaired semantic memory may have difficulty understanding the names of objects, yet have no difficulty recognizing pictures of those same objects (McCarthy and Warrington, 1988; Lauro-Grotto *et al*., 1997*a*). Patients have also been documented who show the opposite pattern: better understanding of words than pictures (McCarthy and Warrington, 1986; Warrington and McCarthy, 1994). This double dissociation has been interpreted (McCarthy and Warrington, 1988; Warrington and McCarthy, 1994) as evidence of separate modality-specific semantic systems. Nevertheless, performance dissociations are rarely absolute. In most cases of semantic dementia, both words and pictorial stimuli are affected, albeit to differing degrees. Moreover, knowledge elicited from pictures and words has been found in some studies to be highly correlated (Lambon Ralph *et al*., 1999), which would not be predicted if that knowledge was stored in separate verbal and visual semantic systems. An opposing view is that there is a unitary semantic storage system, which is itself amodal but accessible from each input modality (Humphreys and Riddoch, 1988; Caramazza *et al*., 1990). Findings of superior comprehension for pictures than words have been ascribed to the more direct mapping between surface form and meaning in the case of pictures (Lambon Ralph and Howard, 2000). For example, the presence of a handle and lip on a jug suggests *a priori* that the jug is an object to be handled and poured rather than something to eat. Pictures contain featural information that provides clues to meaning. Words, by contrast, are arbitrary labels.

Such an explanation holds only for performance differences in favour of pictures. The unitary amodal model would not predict dissociations in the reverse direction. When poorer performance for pictures occurs it has been ascribed to the presence of additional visual processing deficits, arising at a peripheral, pre-semantic level (Humphreys and Riddoch, 1988; Caramazza *et al*., 1990). Thus, it would be assumed that the patients described by McCarthy and Warrington (1986) do not have a pure semantic disorder, but have additional perceptual processing impairments that compromise picture recognition.

Names and faces represent a potentially valuable means of addressing the unitary versus multiple semantic system debate. A person's face, like their name, is arbitrary. There are no intrinsic featural clues to identity that make face recognition inherently easier than names.

Semantic dementia and progressive prosopagnosia

The investigation of famous face recognition as part of a study of semantic memory is particularly topical. Recent years have seen a series of case reports describing patients with progressive prosopagnosia due to focal degenerative disease (Tyrrell *et al*., 1990; Barbarotto *et al*., 1995; Evans *et al*., 1995; Gentileschi *et al*., 1999, 2001; Gainotti *et al*., 2003; Joubert *et al*., 2003). In many, although not all (Joubert *et al*., 2003) cases the anterior, inferolateral parts of the right temporal lobe are particularly affected, areas homologous to those of the left temporal lobe affected in semantic dementia. In such cases the form of prosopagnosia is associative in type. Nevertheless, the precise relationship between progressive prosopagnosia and semantic memory and, by implication, semantic dementia is poorly understood, a point highlighted eloquently by Gainotti *et al*. (2003).

Examination of recognition of famous faces and names in patients with an established circumscribed semantic disorder ought to shed light on the relationship between semantic dementia and progressive prosopagnosia.

Methods

Purpose of study

The primary purpose of the study was to address the relationship between knowledge of famous faces and names in patients with established semantic dementia. An amodal model of semantic memory would predict a systematic relationship between recognition of famous faces and their corresponding name. By contrast, a multiple-semantics model would predict dissociations in name and face recognition. A secondary aim was, by examination of face recognition in patients with an established semantic disorder, to shed light on the relationship between semantic dementia and progressive prosopagnosia.

Studies of semantic dementia typically involve single cases or very small groups of cases, reflecting the rarity of the disorder. We have had the opportunity to report data from a relatively large

Table 1 *Demographic characteristics*

	Semantic dementia	Alzheimer's disease	Control
Number	15	17	30
Male:female	9:6	10:7	16:14
Age at test: mean years (SD)	66 (6)	67 (7)	62 (8)
Illness duration: mean years (SD)	4.4 (2)	4.4 (2)	n/a
MMSE	21 (7)	20 (5)	n/a

consecutive series of 15 patients. Of particular interest is that the cohort includes not only patients with a predominance of atrophy in the left temporal lobe but also some with greater right temporal atrophy. The inclusion of amnesic Alzheimer's disease patients as a reference group was designed to circumvent potential problems in data interpretation arising from control ceiling effects and to ensure that performance in the semantic group indeed reflects their semantic disorder and is not a general product of cognitive decline. The study formed part of an investigation into the neuropsychology of frontotemporal lobar degeneration and was approved by the local ethics committee.

Participants

Semantic dementia

This group comprised 15 patients with semantic dementia, seen in a specialist young-onset dementia unit over a period of 5 years (Table 1). At their initial presentation all patients had complained of problems in naming and comprehension. In 10 (67%) complaints extended also to the recognition of faces and objects, and in three (20%) this was a dominant problem. Patients' day-to-day memory was well preserved, and all patients could find their way around their locality without becoming lost. All were fully independent in activities of daily living. Compulsive behaviours, such as clock-watching, repetitive behavioural routines and preoccupation with a limited set of activities, previously reported to be characteristic of semantic dementia (Snowden *et al*., 2001) were reported in 12 patients (80%). Neurological signs were absent in 12 patients (80%) and limited to mild grasp reflexes in two (14%). In a single patient (7%) slight weakness and wasting of the small muscles of the hand was detected and neurophysiological investigation revealed widespread anterior horn cell disorder compatible with motor neuron disease. A positive family history of dementia in a first degree relative was present in four patients (27%). In all patients neuroimaging revealed atrophy most marked in the temporal lobes, in 10 most marked on the left, in three on the right and in two with no obvious asymmetry. Two of the patients have been the subject of an earlier study and their case histories provided in detail (Snowden and Neary, 2002).

Alzheimer's disease

Seventeen patients with a severe classical amnesia due to Alzheimer's disease served as a reference group (Table 1). All patients had presented with progressive memory impairment in the absence of a history of vascular disease, head injury or alcohol abuse. Patients were physically well at the time of examination and neurological examination was either entirely normal or showed mild rigidity. Patients' Hachinski ischaemia score was <4. All patients had undergone neuroimaging during the course of initial diagnostic investigations, which confirmed the presence of cerebral atrophy, involving the hippocampus, and excluded vascular disease and treatable neurosurgical conditions. All patients had been followed-up for a minimum period of 1 year, and the progressive nature of memory impairment had been documented. Patients with obvious aphasia, perceptual or spatial deficits were excluded to minimize confounding effects on performance. In eight patients there was a family history of Alzheimer's disease. Alzheimer's disease patients were matched to the semantic dementia patients with respect to demographic features and a measure of clinical severity. The two groups did not differ significantly in terms of age ($t = 0.55$, $P = 0.6$), duration of illness ($t = 0.02$, $P = 0.99$) or Mini Mental State Examination (MMSE) scores ($t = 0.75$, $P = 0.5$).

Normal controls

Thirty healthy control subjects (Table 1) were drawn from relatives of patients attending the Cerebral Function Unit young-onset dementia clinic and were predominantly spouses of patients in this study. No control had a history of neurological disease, vascular disease, head injury or alcohol abuse. All were well, were not under treatment for a major illness and had no cognitive complaints. The rationale for selecting patients' spouses was to control as far as possible for socio-economic background and likely prior exposure to the famous faces and names. The normal controls did not differ significantly from the semantic group in terms of age ($t = 1.67$, $P = 0.09$), although they were slightly younger than the Alzheimer's disease group ($t = 2.27$, $P = 0.03$).

Background neuropsychology

Background neuropsychological test data for the semantic and Alzheimer's disease groups are shown in Table 2. The patients with semantic dementia were highly impaired on tests of naming and single word comprehension. They performed significantly worse than the Alzheimer's disease patients on the difficult Graded Naming test (McKenna and Warrington, 1983) ($t = 8.72$, $P < 0.0001$), an easy locally developed picture naming test ($t = 6.36$, $P < 0.0001$) and a locally developed forced-choice word-picture matching test ($t = 3.11$, $P = 0.004$). Mean performance was within normal limits in both semantic and Alzheimer's disease groups on subtests of the Visual Object and Space Perception (VOSP) Battery (Warrington and James, 1991), with the exception that the semantic group was impaired on the Silhouettes sub-test, which requires recognition of pictures of animals and objects. Scores were significantly worse than in the Alzheimer's disease group ($t = 6.53$, $P < 0.001$). By contrast, the semantic dementia group performed better than the Alzheimer's disease group on spatial sub-tests: position discrimination ($t = 2.46$, $P = 0.02$), number location ($t = 2.40$, $P = 0.02$) and cube analysis ($t = 2.10$, $P = 0.04$). Both semantic and Alzheimer's disease groups

Table 2 *Background neuropsychological data*

Test	Mean scores (SD)		
	Semantic dementia	Alzheimer's disease	Normal mean or *5% cut-off*
Graded Naming Test/30	**0**	18 (8)	23 (4)
'Easy' Naming test/40	**16 (13)**	37 (3)	39 (1)
'Easy' word-picture match/40 (chance = 10)	**30 (12)**	40 (0)	40 (0)
Pyramids and Palm Trees: words/52 (chance = 26)	**36 (8)**	–	*47*
Pyramids and Palm Trees: pictures/52 (chance = 26)	**40 (6)**	–	*47*
VOSP – screening test/20	20 (0)	20 (0)	*15*
VOSP – incomplete letters/20	17 (6)	19 (1)	*16*
VOSP – silhouettes/30	**6 (5)**	18 (4)	*15*
VOSP – object decision/20	15 (3)	17 (2)	*14*
VOSP – dot counting/10	10 (0)	10 (0)	8
VOSP – position discrimination/20	20 (0)	18 (2)	18
VOSP – number location/10	10 (0)	8 (2)	7
VOSP – cube analysis/10	9 (1)	8 (2)	6
Picture recall/20	**2 (3)**	**2 (2)**	10 (2)
Picture recognition/20 (chance = 5)	16 (3)	**11 (4)**	19 (1)
Orientation/10	9 (1)	**4 (3)**	*9*

Bold values indicate impaired performance.

performed very poorly on a test of picture recall, which makes verbal demands. However, in a four-choice recognition version of the task the semantic group performed significantly better than the Alzheimer's disease group (t = 3.56, P = 0.001). They also performed better on the orientation questions from the MMSE (t = 4.84, $P < 0.0001$) and, in particular, on the temporal orientation items (t = 6.97, $P < 0.0001$). The semantic dementia patients were impaired on both the word and picture versions of the Pyramids and Palm Trees test (Howard and Patterson, 1992). These findings show that semantic patients perform worse than amnesic Alzheimer's disease patients on tests that make semantic demands, but better on other cognitive tasks.

Famous face and famous name test

Famous faces: materials

The test materials consisted of photographs of 75 famous people alive during the subjects' lifetime. Some were contemporary individuals (e.g. Tony Blair, David Beckham) whereas others were from the more distant past (e.g. Winston Churchill, Diana Dors). For each famous face a non-famous face was selected of the same sex and of a similar age, matched for general physical appearance to minimize the possibility that one face should look inherently more 'famous' than the other. All faces were presented in an identical format as black and white portrait photographs of 11 × 13 cm dimension. Each famous face with its non-famous matched pair was presented side by side in an A4-sized folder. The positioning of the famous face in the left and right positions was counter-balanced.

Famous names: materials

The test materials consisted of the names of the same 75 famous people shown in the famous faces task. For each famous name a non-famous name was selected, of the same sex and matched for name characteristics. Hence, a diminutive (Ted) was matched with another short-form (Joe), a foreign name (Portillo) was matched with another foreign name (Romero), and a first name–surname combination with assonance (Mo Mowlam) was matched with a first name–surname combination also with similar sounds (Di Dillon). Each famous name with its non-famous counterpart was presented side by side in an A4-sized folder. The positioning of the famous name in left and right positions was counter-balanced.

Procedure

The famous faces were presented first. Each subject was shown each photograph pair in turn and asked to indicate the famous person and to guess if they did not know. Guesses were recorded. The subject was then asked to provide identifying information and to name the face if possible. Names were considered correct only if both first name and surname was given. No feedback was given. In the case of identifying information, a lenient criterion was accepted, to compensate for the severely limited descriptive vocabulary available to patients with semantic dementia. A response of the type 'he's the top man' in response to a face of Tony Blair would be regarded as correct, as would the response 'the president down there in London', since the response conveys correct identification even though terminological usage is incorrect. Generic occupational terms were also accepted for a film actor, provided that they clearly demonstrated contextual recognition. An emphatic response such as 'He's in all the old films' would be accepted, whereas a vague general response such as 'Is he on television?' would not. No feedback was given and, in particular, on no occasion was the name corresponding to the famous face given by the examiner.

After a short delay the subject was shown each name pair, and asked to point to the one of the two that was famous and to guess if they did not know. Guesses were recorded. The subject was then asked to provide identifying information about the name. A lenient criterion, identical to that used for the face task, was adopted for designation of a correct response. The subject was not informed that the names corresponded to the people shown in the face test.

Six of the 15 semantic dementia patients were administered the Famous Face and Name tasks a second time after 1 year to examine response consistency and change in performance over time.

Table 3 *Summary data for the famous face and name tasks*

Task	SD	Alzheimer's disease	Control
	Mean (SD)	Mean(SD)	Mean (SD)
Naming famous faces	2 (2)	13 (10)	43 (12)
Identification of famous faces	11 (13)	35 (17)	63 (10)
Identification of famous names	10 (12)	55 (16)	71 (4)
Forced-choice familiarity for faces	56 (13)	66 (6)	71 (4)
Forced-choice familiarity for names	51 (12)	71 (4)	74 (1)
Explicit report of 'knowing' face	32 (23)	58 (11)	69 (5)
Explicit report of 'knowing'name	27 (24)	70 (5)	74 (1)

Maximum score for all tasks = 75.

Statistical analyses

One-way and two-way repeated measures analyses of variance were carried out, with group (semantic versus Alzheimer's disease versus control) as the between subjects factor and test modality (faces versus names) the within subjects factor. Main and interaction effects were further examined using planned *t*-test comparisons (*a priori* tests). Correlative analyses used Pearson's statistic and contingency coefficients. McNemar tests examined the significance of changes in performance over time and as a function of test modality.

Results

Mean scores for each group for each testing condition are shown in Table 3.

Naming famous faces

There was a significant main effect of group [$F(2,59) = 94.47$, $P < 0.0001$]. The semantic dementia patients named fewer faces than the Alzheimer's disease patients ($t = 3.81$, $P = 0.001$), who in turn named fewer faces than controls ($t = 8.49$, $P < 0.0001$).

Identification of famous faces and names

There was a highly significant main effect of group [$F(2,59) = 124.68$, $P < 0.0001$]. Semantic dementia patients identified fewer names/faces than the Alzheimer's disease patients ($t = 6.55$, $P < 0.0001$), who in turn identified fewer than controls ($t = 6.90$, $P < 0.0001$). There was a highly significant main effect of test material [$F(1,59) = 61.00$, $P < 0.0001$]. Famous names were identified significantly better than famous faces. There was also a significant group × test material interaction [$F(2,59) = 21.71$, $P < 0.0001$]. Both the Alzheimer's disease and control groups performed better on the name than the face identification task ($t = 9.64$, $P < 0.0001$ and $t = 5.25$, $P < 0.0001$ for the two groups, respectively). By contrast, the semantic dementia group had numerically better scores for famous faces than famous names, although the difference did not reach statistical significance.

Error analysis

All errors in the Alzheimer's disease and control groups constituted 'don't know' responses. There were no instances of misattribution of identity. In the semantic group, most patients similarly made exclusively 'don't know' responses for unidentified stimuli. Nevertheless, one semantic patient misidentified 13 faces (17%) and three names (4%). In two instances misattributions were of semantically related individuals (e.g. Margaret Thatcher identified as the Queen). However, in 14 instances famous people were ascribed the identity of a personal acquaintance (e.g. Tony Blair: 'he's down at the engineering works. I saw him last week'; Hillary Clinton: 'she's in the typing pool at the office'; Elvis Presley: 'I know him. He's in the building trade'; John Prescott: 'He goes to our church. He's involved in church activities').

Familiarity of famous faces and names

Forced-choice familiarity judgements

There was a highly significant main effect of group [$F(2,59) = 54.43$, $P < 0.0001$]. Semantic dementia patients were significantly more impaired in judging familiarity than the Alzheimer's disease patients ($t = 5.33$, $P < 0.0001$), who in turn performed significantly worse than controls ($t = 4.81$, $P < 0.0001$). There was no main effect of stimulus modality [$F(1,59) = 1.14$, $P = 0.29$]. However, there was a significant group × modality interaction effect [$F(2,59) = 8.55$, $P = 0.001$]. Both Alzheimer's disease patients and controls made significantly better familiarity judgements for names than for faces (Alzheimer's disease: $t = 3.27$, $P = 0.005$; controls: $t = 5.172$, $P < 0.0001$). In the semantic dementia group numerical scores were in the reverse direction, although this difference did not reach statistical significance.

Explicit reports of familiarity ('feeling of knowing')

Forced-choice familiarity judgement scores combine correct responses based on genuine feelings of familiarity and correct guesses. An analysis based only on explicit reports of 'knowing' the name or face revealed a highly significant

Table 4 *Summary data for the two semantic sub-groups*

	Left > right temporal lobe atrophy	Right > left temporal lobe atrophy
	Mean (SD)	Mean (SD)
Naming famous faces/75	3 (2)	2 (3)
Identification of famous faces	15 (15)	5 (5)
Identification of famous names	9 (14)	17 (18)
Forced-choice familiarity for faces	60 (11)	55 (17)
Forced-choice familiarity for names	49 (11)	63 (13)
Explicit report of 'knowing' face	39 (23)	17 (15)
Explicit report of 'knowing'name	24 (21)	42 (37)

Maximum score for all tasks = 75.

group effect [$F(2,59) = 1477.9$, $P < 0.0001$]. The semantic patients reported explicit familiarity for fewer items than both the Alzheimer's disease and controls groups ($P < 0.0001$). Subjective reports of familiarity did not differ in the Alzheimer's disease and control groups ($P = 0.09$). There was a small effect of modality [$F(1,59) = 4.49$, $P < 0.04$], reflecting more explicit reports of familiarity for names than faces, and a larger group × modality interaction effect [$F(2,59) = 5.97$, $P = 0.004$]. Whereas semantic patients reported more explicit feelings of familiarity for faces than names the Alzheimer's disease and control groups showed the reverse finding.

Relationship between forced-choice familiarity judgements and explicit reports of familiarity 'feeling of knowing'

In a two-choice forced recognition test 'guesses' based on a lack of explicit feeling of familiarity should yield 50% correct and 50% incorrect responses. We selected items that subjects reported to be 'guesses', and carried out a binomial test on these responses to determine whether correct selection differed from chance.

Correct 'guesses' did not differ from chance in any subject in the control group and in only one Alzheimer's disease patient for famous faces ($P < 0.0001$). However, in six of the 15 semantic dementia patients (40%) correct 'guesses' occurred significantly more often than expected by chance for either names or faces or both (names: Patient 5, $P = 0.009$, Patient 15, $P = 0.006$; faces: Patient 3, $P = 0.03$, Patient 5, $P = 0.001$, Patient 9, $P = 0.04$, Patient 10, $P = 0.004$, Patient 11, $P = 0.02$). Patients exhibiting this qualitative performance feature were all in the middle range of severity with respect to the rest of the group in terms of their name or face score (ranking 4–9).

Face and name performance and distribution of temporal lobe atrophy

The semantic dementia patients were divided into those patients in whom there was a predominance of atrophy in the left temporal lobe (10 patients) and those in whom right temporal atrophy predominated (three patients). Two patients, in whom the distribution of atrophy was reported to be bilateral and symmetrical, were excluded from the analysis. Judgement of asymmetry was based on neuroradiological reports and inspection by an independent neurologist who was blind to the neuropsychological test results. The two sub-groups (left > right atrophy and right > left atrophy) were compared with respect to their performance on name and face tasks (Table 4).

Naming famous faces

There was no difference in naming scores in the two sub-groups [$F(1,11) = 0.21$, $P = 0.66$].

Identification of famous faces and names

There was no main effect of sub-group [$F(1,11) = 0.007$, $P = 0.94$] or modality [$F(1,11) = 1.84$, $P = 0.2$], showing that the sub-groups were matched in overall level of performance, and there was no overall superiority for names or faces. However, there was a significant interaction effect [$F(1,11) = 12.02$, $P = 0.005$]. The left > right atrophy group identified faces better than names ($t = 3.09$, $P = 0.01$), whereas those with right > left atrophy showed the converse pattern (Fig. 1A). The latter paired-comparison did not reach statistical significance because of the very small group size ($n = 3$).

Familiarity of famous faces and names

There was no significant main effect of sub-group [$F(1,11) = 0.36$, $P = 0.56$] or modality [$F(1,11) = 0.12$, $P = 0.74$]. There was, however, a significant sub-group × modality interaction [$F(1,11) = 7.58$ $P = 0.02$]. Whereas the left > right atrophy sub-group showed superior face familiarity judgements ($t = 3.09$, $P = 0.01$), the right > left atrophy group showed numerically superior name familiarity performance. The latter comparison did not reach statistical significance because of the very small sample size ($n = 3$).

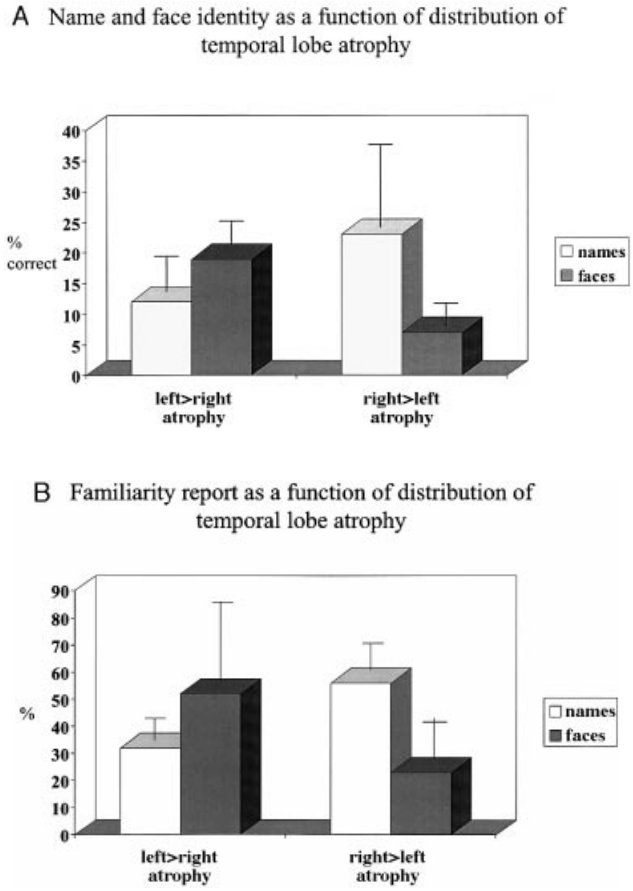

Fig. 1 Identification performance (**A**) and familiarity reports (**B**) for names and faces in semantic dementia as a function of distribution of temporal lobe atrophy. The figures show mean percentage scores and standard errors.

When familiarity was based on explicit 'feeling of knowing' rather than forced-choice test scores the findings were similar. There was no overall sub-group effect [$F(1,11) = 0.02$, $P = 0.9$] nor modality effect [$F(1,11) = 0.58$], but there was a significant interaction effect [$F(1,11) = 8.19$, $P = 0.02$]. Patients with left > right atrophy reported familiarity for more faces than names ($t = 2.2$, $P = 0.05$), whereas patients with right > left atrophy showed the converse pattern (Fig. 1B). Again, the latter paired-comparison did not reach statistical significance because of the small sample size. Examination of individual test scores showed a direct one-to-one correspondence between numerical superiority of performance for names or faces and left or right emphasis of atrophy (contingency coefficient = 0.71, $P < 0.0001$).

Interrelationship between face and name performance

The ability to identity famous faces and names was weakly correlated in controls ($r = 0.44$, $P = 0.02$) but showed a stronger correlation in the Alzheimer's disease ($r = 0.87$, $t < 0.001$) and semantic ($r = 0.77$, $t = 0.001$) groups. Familiarity judgements based on explicit report of 'knowing' revealed a high correlation in the control group ($r = 0.60$, $t < 0.0001$), to a slightly lesser extent in the Alzheimer's disease group ($r = 0.58$, $P = 0.01$), but were uncorrelated in the semantic dementia group ($r = 0.44$, $t = 0.10$).

Intercorrelations in the semantic dementia group between face and name performance and background tests of semantic memory

Scores on the famous face and name tasks did not correlate uniformly with scores on the background tests of semantic memory (Table 5). Name tasks were highly correlated with semantic tests that make strong verbal demands (picture naming) or are exclusively verbal (word version of the Pyramids and Palm Trees test) but were more weakly correlated with tests that make comparable visual–verbal demands (word–picture matching) or are exclusively visual (picture version of the Pyramids and Palm Trees test). Face tests showed weaker and less consistent correlations with the background semantic memory measures than name tests.

Relative superiority of identification performance for famous faces compared with names was associated with superior performance on the picture compared with the word versions of the Pyramids and Palm Trees test (contingency coefficient 0.49, $P = 0.03$). Scores on the word and picture versions of the Pyramids and Palm Trees test were not themselves correlated ($r = 0.45$, $P = 0.23$). The Graded Naming test did not yield significant correlations because of floor level scores in all patients.

Item concordance for faces and names in semantic dementia patients

The semantic group includes a wide range of illness severity, with some scores approaching normal levels and others at floor level. Significant name–face correlations might potentially occur because of these very wide overall patient differences. To eliminate effects of overall severity, individual within-subject analyses were carried out for each semantic dementia patient, examining item-by-item correspondence for names and faces.

There was a statistically significant ($P < 0.05$) correspondence between explicit familiarity reports for name–face pairs in only five of 11 (45%) patients. (Four patients reported no names to be familiar so a meaningful correlation was not possible.) There was a significant correlation ($P < 0.01$) between identification performance for faces and their corresponding names in all 10 patients in whom a correlation could be computed.

Notwithstanding the partial correspondence between responses for name–face pairs there was, as noted above, a numerical superiority in test scores for faces in all patients with left > right atrophy and a superiority for names in all

Table 5 *Correlation in semantic dementia between famous name and face performance and background tests of semantic memory*

	Picture naming		Word-picture match		Pyramids and Palm Trees (words)		Pyramids and Palm Trees (pictures)	
	r	*P*	*r*	*P*	*r*	*P*	*r*	*P*
Face naming	0.83	0.0001 ***	0.61	0.02 *	0.71	0.02 *	0.78	0.01 *
Name identification	0.67	0.007 **	0.41	0.13 n.s.	0.81	0.005 **	0.22	0.57 n.s.
Name familiarity	0.77	0.001 **	0.49	0.07 n.s.	0.90	0.000 ***	0.26	0.50 n.s.
'Knows' name	0.74	0.002 **	0.58	0.02 *	0.88	0.001 **	0.18	0.65 n.s.
Face identification	0.60	0.02 *	0.51	0.05 *	0.60	0.07 n.s.	0.60	0.09 n.s.
Face familiarity	0.51	0.05 *	0.46	0.09 n.s.	0.82	0.004 **	0.54	0.14 n.s.
'Knows' face	0.27	0.33 n.s.	0.25	0.38 n.s.	0.71	0.02 *	0.62	0.07 n.s.

***Significant at $P < 0.001$; **significant at $P < 0.01$; *significant at $P < 0.05$.

patients with right > left atrophy. The difference between familiarity judgement performance for names and their corresponding faces reached statistical significance (McNemar test, $P < 0.05$ to $P < 0.0001$) in 50% of those patients in whom a meaningful measurement of change could be computed (i.e. performance above chance/floor level). In the case of identification responses there was a significant difference in performance (McNemar test, $P < 0.05$ to $P < 0.0001$) for names compared with faces in 56% of patients in whom a measurement of change could be computed.

Test–retest performance in semantic dementia patients

Item concordance

Test–retest comparisons showed a strong item-by-item concordance (at least $P < 0.001$) for explicit familiarity reports as well as name and face identification responses in all patients. These findings confirm that the availability of semantic information about a person is stable, and is not subject to variable access.

Change over time

In keeping with the progressive nature of the patients' disorder it was anticipated that detectable decline in scores should be detected over a 1-year interval. The presence of significant change, as determined by the McNemar test, varied across the patient group. Two patients showed no significant change over 1 year on any measure. In three patients decline occurred on both name and face tasks: in one in explicit report of familiarity for names ($\chi^2 = 11.5$, $P < 0.001$) and faces ($\chi^2 = 8.7$, $P = 0.002$), name identification ($\chi^2 = 19.9$ $P < 0.001$) and face identification ($\chi^2 = 4.3$, $P = 0.04$); in another name ($\chi^2 = 25.3$, $P < 0.001$) and face identification ($\chi^2 = 21.3$, $P < 0.001$) and, in the third, in explicit report of familiarity for names ($\chi^2 = 11.4$, $P < 0.001$), and in identification of faces ($\chi^2 = 8.1$, $P = 0.002$). In one patient deterioration was detected only in one modality: in explicit report of familiarity for faces ($\chi^2 = 4.0$, $P = 0.04$), as well as face identification ($\chi^2 = 4.2$, $P = 0.03$).

Discussion

Semantic dementia patients performed significantly worse than amnesic Alzheimer's disease patients and controls on the famous face and name tasks, confirming the sensitivity of such tasks to breakdown in semantic memory. The semantic disorder affected patients' feeling of familiarity as well as their ability to identify names and faces.

Interestingly some semantic dementia patients reported a lack of familiarity even when they achieved above-chance performance on a forced-choice familiarity judgement test. Inevitably, the interpretation of subjective report data can be problematic because individuals differ in their threshold for reporting familiarity. However, such individual differences ought to apply equally to all groups, yet a disparity between subjective sense of familiarity and forced-choice judgements occurred in 47% of the semantic group, only 6% of the Alzheimer's disease group and 0% of the control group. Moreover, the disparity was always in the same direction: an under-report rather than over-report of familiarity. Furthermore, the semantic patients in whom the phenomenon occurred were all middle-ranking (clustered together) in terms of their performance scores, suggesting a commonality with respect to their level of semantic disorder. Finally, in all but one patient, the disparity between subjective report and actual performance was observed for names or faces only. A cautious 'personal style' ought to apply equally to test materials regardless of input modality. Our interpretation of these data is that semantic loss is not all or none. Just as a patient may have sufficient information about a name or face to support familiarity judgements but not identification or naming, so too there may be differences with respect to familiarity itself. At one extreme, patients may have sufficient residual semantic information to support forced-choice judgements based on an overt feeling of familiarity. At the other extreme, information may be lost to such an extent that the patient has no sense of familiarity and this is reflected in an inability to distinguish previously familiar from unfamiliar stimuli. A putative intermediate position is that residual information is degraded to an extent that it no longer elicits explicit subjective feelings of familiarity and yet is sufficient to support implicit forced-choice familiarity judgements. The

patient selects the correct item more often than would be expected by chance and yet has no confidence in his responses because of the very weak information upon which those judgements are made. A similar threshold argument has been proposed by De Haan *et al*. (1992) to account for covert recognition in patients with impaired overt face recognition. McNeil and Warrington (1991) also reported above-chance level on a forced-choice familiarity task in prosopagnosic patients. Evans *et al*. (1995) too reported, in their patient with progressive prosopagnosia, better than chance performance on forced-choice familiarity judgements of faces despite the patient's impression that he was guessing.

Misattribution of names and faces

In Alzheimer's disease and controls there were no instances of misattribution. Faces and names were either clearly known or not known. In the semantic group too examples of misattribution were rare. However, one patient ascribed three famous names, 12 famous faces and one non-famous face the identity of personal acquaintances. Vuilleumier *et al*. (2003) described a patient, with a unilateral right inferior temporal-occipital lesion who experienced hyperfamiliarity for unknown faces. The present case appears rather different. There was no general increase in level of reported familiarity. Indeed, he reported most previously familiar faces as unfamiliar and he correctly rejected unknown faces as unfamiliar. Our interpretation is that, for some faces, he experiences a sense of familiarity. He feels that he ought to know the person. However, he has a very severe loss of semantic knowledge that has effectively narrowed his conceptual world to his immediate environment and daily experience. He tries to map his sense of familiarity on to the only context available to him: his own personal world.

Modality effects: one or more semantic memory systems?

The findings of the present study are problematic for accounts of semantic memory as a unitary, amodal system (Humphreys and Riddoch, 1988; Caramazza *et al*., 1990). The semantic dementia patients as a group showed the typically reported pattern of superior performance for visual than verbal information. However, in contrast to studies using pictures and words, this cannot be attributed to a putative closer mapping between visual information and meaning. It would be reasonable to assume that the name of a famous person (e.g. Elvis Presley, Margaret Thatcher) is more intimately linked to their identity than is their face. Facial characteristics change over time, whereas a person's name remains constant. Moreover, it would be difficult to envisage media exposure or discussion of the person without recourse to their name. In line with this argument, both amnesic Alzheimer's disease patients and normal controls performed significantly better on the name than the face tasks, a finding consistent with reports of others (Young *et al*., 1986*a*) that name recognition is easier than face recognition. The semantic group is thus showing greater impairment on what is ostensibly the easier task.

An even more compelling finding is that face superiority is not an invariable finding in the semantic group. There is a clear relationship between the direction of performance (names better than faces or faces better than names) and the distribution of atrophy. Patients with predominant left temporal atrophy perform better with faces; patients with predominant right temporal atrophy perform better with names. These left and right predominant sub-groups do not differ in terms of demographic variables, duration of illness or performance on baseline neuropsychological tests, including standard perceptual tasks. There are no grounds, therefore, for attributing coincidental cognitive deficits to one semantic sub-group and not the other. Indeed, the distribution of atrophy on structural brain imaging is uniform (affecting the anterior inferolateral parts of the temporal lobes), the only difference being the relative emphasis of left or right. The most parsimonious explanation is that there are real differences in the nature of patients' semantic memory loss, which are directly related to the distribution of the atrophy within the left and right temporal lobes.

The data complement other reports of hemispheric differences in the recognition of names and faces. Eslinger *et al*. (1996) described a patient with left temporal damage who was unable to access information about famous people from their name but could do so from their face, and a second patient with predominantly right hemisphere damage who showed the reverse pattern. Verstichel *et al*. (1996) reported a selective deficit in name recognition associated with left temporal damage. Haslam *et al*. (2001) reported a patient with right temporal damage due to herpes simplex encephalitis who accessed more information about famous people from their name than their face.

Do the data therefore support the existence of separate semantic systems for verbal and non-verbal information, as proposed by McCarthy and Warrington (1988)? Some aspects of the data are problematic for the modality-specific account too. In both Alzheimer's disease and semantic groups the ability to identify a famous person from their name and from their face was significantly correlated. It has been argued (Lambon Ralph *et al*., 1999) that separate semantic systems would not predict such correlations. A possible counter-argument might be that the semantic group includes a wide range of levels of severity; at one extreme, patients recognize no famous people and, at the other, performance is only mildly impaired. Such wide differences might yield general positive correlations across name and face tasks simply by virtue of the overall magnitude of the degenerative process, which affects both hemispheres. The examination of item-by-item correspondence in the semantic dementia group is potentially informative because it overcomes the confounding factor of disease severity by addressing within- rather than between-subject performance. Significant item-by-item correlations were present both for judgements of familiarity

and identification responses. The interpretation is not wholly straightforward. Famous people are not equally famous. They receive differing degrees of media exposure and vary in their semantic salience (compare Margaret Thatcher and Alec Douglas-Home as two former British Prime Ministers). General positive name/face correlations might potentially arise as a result of differences in level of stimulus 'difficulty'. The influence of overall semantic salience in the present study cannot be firmly excluded. Nevertheless, explicit familiarity reports are of interest because responses are well above floor level but below ceiling, so are less likely to yield spurious correlations. Explicit familiarity reports for names and their corresponding face were correlated in most but not all patients. It is instructive to note too that Lambon Ralph and Howard (2000) have demonstrated significant item consistency even when such factors as overall familiarity or difficulty were controlled for.

The existence of such correlations presents some difficulty for an account of semantic memory in terms of separate semantic systems. Nevertheless, the fact that name and face performance is not invariably correlated is equally problematic for the unitary, amodal model. Moreover, in those patients who were re-assessed after 1 year there was not always a comparable decline in name and face performance. Furthermore, performance across semantic tasks was not uniformly correlated. Famous name performance was strongly correlated with verbal semantic measures (picture naming, and Pyramids and Palm Trees word test) but less with a semantic task that makes comparable visual–verbal demands (word–picture matching) and even less with a visual semantic task (Pyramids and Palm Trees picture test). Performance on the word and picture versions of the Pyramids and Palm Trees test was not itself inter-correlated, yet there was a significant association between word or face superiority on the Famous Names and Faces tests and better performance, respectively, on the word or picture version of the Pyramids and Palm Trees test. These factors, together with the demonstration of a double dissociation in name and face performance in patients with predominant left and right temporal lobe atrophy, argue strongly against a unitary amodal account of semantic memory. How then can the data be reconciled?

The multiple-system account, in its original form (Warrington and McCarthy, 1988), lacks parsimony as it requires reduplication of information within separate modality-specific systems. A less strong view (Lauro-Grotto *et al*., 1997*b*) is that the semantic system is a multimodal network, in which different areas are accessed by each modality and store modality specific information. Under normal circumstances it is assumed that the various components of the net are inter-connected, allowing retrieval of the entire representation from any input channel. However, in pathological conditions one or more components of the net can be preferentially damaged, giving rise to dissociations in performance.

We regard our data as consistent with the notion of semantic knowledge as an integrated, multimodal network of knowledge, with the left temporal lobe being particularly important for verbal information and the right temporal lobe visual information. Such a model predicts that performance should be influenced both by the overall extent and the locus of damage. Extensive damage, resulting from severe temporal lobe atrophy, would have an overall more disruptive effect on the network than mild damage, hence the general overall correlation in performance across tasks. However, dissociations in performance would also be possible, reflecting the site of damage within the network. It is of interest that, in the present cohort of semantic dementia patients, the magnitude of the difference in performance for names and faces is variable. Such gradations in extent of dissociation can readily be accommodated by a multimodal network model, which assumes that disruption to different parts of the net is relative rather than absolute.

We do not propose that the anterior temporal lobes 'store' concepts. Rather, in parallel with the notion 'convergence zones' put forward by Damasio and colleagues (Damasio and Damasio, 1994; Tranel *et al*., 1997) and 'transmodal areas' advanced by Mesulam (1998) we assume they have a integrative role in binding together components of information. Critically, however, we assume a division in neural architecture, with the anterior regions of the right temporal lobe specialized for visual information and the left for verbal. This position differs somewhat from that proposed by Lambon Ralph *et al*. (2001). Those authors found disproportionate impairment in naming relative to comprehension in semantic dementia patients with predominantly left temporal atrophy compared with those with predominantly right temporal atrophy. The authors interpreted their findings within the framework of a unitary model of semantics in which conceptual knowledge is distributed across both temporal lobes and there is no division of cognitive/neural architecture. They ascribed the disproportionate naming impairment in patients with left temporal atrophy to stronger connections between semantic units on the left with left-lateralized phonological representations, i.e. they assume no differentiation within the semantic units themselves. Nevertheless, their data might plausibly also be explained in modality-specific terms. Naming and comprehension were measured by picture naming and word–picture matching tasks, both involving visual and verbal modalities. A putative impairment in visual semantic information, arising from right hemisphere pathology, would compromise picture recognition, which would be expected to affect comprehension and naming performance in an essentially uniform way. By contrast, a disorder of verbal semantic information, associated with left hemisphere pathology, would be expected to impair naming more than comprehension because the presence of partial or degraded information may be sufficient to support forced-choice matching performance but not naming.

A principle and fundamental conceptual difference between the postulated multimodal account of semantic

knowledge that we wish to advance and an amodal model of semantic memory is, in our own view, that in the former, modality-specific information forms part of the body of information that constitutes semantic knowledge. By contrast, in the latter it is merely the vehicle by which semantic knowledge is accessed. In a multimodal account the visual appearance of a person's face is part of network of semantic information that a person has about a person. There is no higher order level. In the amodal account, visual appearance is merely one channel through which abstracted semantic knowledge about a person can be activated. There is a superordinate level that constitutes semantics. This conceptual distinction has a bearing on contemporary models of face recognition.

Implications for models of face recognition

The influential model of face recognition of Bruce and Young (1986*b*) distinguishes four levels of analysis. Structural encoding is the most elementary level and is the process by which facial percepts are formed. This leads to activation of 'face recognition units' (FRUs), which correspond to the stored descriptions of known faces. These face recognition units provide a link between structural encoding of the face's appearance and 'person identity nodes' (PINs), which provide access to stored information about a person (semantics). The final stage of the process is name generation.

All the patients in our study have a circumscribed semantic disorder and we ascribe impairments in face recognition to a semantic level. In terms of the model the deficit would lie at the level of PINs. However, an assumption of the model is that deficits occurring at this level should be cross-modal. Impaired person knowledge should lead to impaired recognition both when the stimulus is a face and a name. Recent modifications of the original Bruce and Young model (Burton and Bruce, 1993) make this prediction even more explicit. Modality specific units, required to identify a person's name or face as familiar, are combined in the person identity nodes, allowing recognition of a particular person, and through this route semantic knowledge is activated.

The model assumes that there is no differentiation between modalities once the PINs or semantic level of processing is reached. It presupposes a higher-order semantic system that is independent of modality. Thus, semantic information available via the face route should also be available via the name route. The model would interpret impairments in recognition arising from one modality, but not another as inevitably arising at an earlier pre-semantic stage of processing, at the level of FRU or in the interface between PINs and FRUs. Indeed, it is precisely this interpretation that was proposed to explain the initial selective prosopagnosia described by Evans *et al*. (1995). Our own findings, as well as those of others (Eslinger *et al*., 1996; Haslam *et al*., 2001), demonstrating dissociations in name and face performance arising at a semantic level, provide a challenge for the model as it currently stands.

Relationship between semantic dementia and progressive prosopagnosia

Progressive prosopagnosia (Tyrrell *et al*., 1990; Barbarotto *et al*., 1995; Evans *et al*., 1995; Gentileschi *et al*., 1999, 2001; Gainotti *et al*., 2003; Joubert *et al*., 2003) has typically been described as a syndrome distinct from that of semantic dementia. There are several reasons why this should be so. First, semantic memory has traditionally been seen as inherently tied to language and the province of the left temporal lobe. Functional imaging studies have provided support for such anatomical lateralization (Demonet *et al*., 1992; Vandenberghe *et al*., 1996), albeit using verbally based tasks. Secondly, the preponderance of patients with semantic dementia who exhibit greater atrophy of the left than the right temporal lobe has further emphasized the critical importance of the left temporal lobe for semantic memory (Hodges *et al*., 1992, 1998). Thirdly, the preconception of semantic memory as an amodal system of knowledge precludes the existence of a central semantic impairment confined to one modality.

Nevertheless, Gainotti *et al*. (2003) has pointed out that, in at least some of the reported cases of progressive prosopagnosia (Barbarotto *et al*., 1995; Evans *et al*., 1995; Gentileschi *et al*., 1999, 2001), the recognition disorder is not confined to face recognition, but rather represents a cross-modal impairment of people knowledge, i.e. a disorder of semantics. The patient reported by Evans *et al*. (1995) initially showed a relatively selective prosopagnosia, but on follow-up had progressed to involve loss of knowledge about people independent of modality of access. The initial deficit was ascribed, in accordance with the Bruce and Young model, to a pre-semantic stage of processing, whereas on follow-up the disorder was assumed to affect semantics. The inference was that there had been a spread of pathology in a forwards direction. We would postulate an alternative interpretation. The disorder from the outset represents a disorder of semantics, but is initially confined to a specific component of semantic knowledge: knowledge of faces, and progresses to involve more widespread loss of semantic information. Anatomically we would assume that the degenerative process is spreading, not from back to front, but from right to left.

In semantic dementia, emphasis on the importance of the left temporal lobe has arisen because most reported cases show greater left temporal atrophy than right. However, atrophy is invariably bilateral, albeit often asymmetrically distributed. Moreover, the predominant atrophy is not always left-sided, as is the case in some patients in the present study. It is instructive that the anterior, inferolateral part of the right temporal lobe that is atrophied in many patients with progressive prosopagnosia is precisely the same region atrophied in semantic dementia. We would argue that progressive prosopagnosia is one of the presentations of semantic dementia. In patients with predominantly left-sided temporal degeneration the disorder may initially be manifest as a problem with naming and understanding words, whereas in patients with predominant right-sided atrophy the semantic

disorder may manifest initially as a disorder of face recognition. In both cases, the degenerative process ultimately leads to a multimodal semantic loss.

The question inevitably arises why left-sided presentations are more common than right-sided presentations. Perhaps one contributory factor is that problems in language are more immediately evident and more troublesome than problems in face processing and so more likely to come to light. In the present series, for example, the patients with predominant right temporal lobe atrophy had come to medical attention only when problems with language had begun to emerge. Yet in all cases relatives acknowledged that the patients' problem with face recognition was the outstanding and earliest feature of their disorder.

In conclusion, the study demonstrates a dissociation between face and name knowledge in patients with semantic dementia, reflecting differential involvement of the right and left anterior temporal lobes. We attribute the dissociation to differential disruption to components of a multimodal network by which semantic knowledge is cerebrally represented.

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