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Relative category-specific preservation in semantic dementia? Evidence from 35 cases

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A R T I C L E I N F O

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A B S T R A C T

Category-specific deficits have rarely been reported in semantic dementia (SD). To our knowledge, only four previous studies have documented category-specific deficits, and these have focused on the living versus non-living things contrast rather than on more fine-grained semantic categories. This study aimed to determine whether a category-specific effect could be highlighted by a semantic sorting task administered to 35 SD patients once at baseline and again after 2 years and to 10 Alzheimer's disease patients (AD). We found a relative preservation of fruit and vegetables only in SD.

This relative preservation of fruit and vegetables could be considered with regard to the importance of color knowledge in their discrimination. Indeed, color knowledge retrieval is known to depend on the left posterior fusiform gyrus which is relatively spared in SD. Finally, according to predictions of semantic memory models, our findings best fitted the Devlin and Gonnerman's computational account.

1. Introduction

Semantic memory is currently defined as a system where general knowledge (or "conceptual knowledge") about words, living and nonliving entities, people, public events and places, is stored in the form of symbolic representations (Tulving, 1972). The notion of semantic memory first appeared in the 1960s (Quillian, 1966), in influential cognitive psychology works on artificial intelligence, with an emphasis on language abilities. Evidence from neuropsychological studies with brain-injured patients subsequently raised questions as to whether semantic memory consists of a unitary system, or whether multiple systems are required. Should it be regarded as an amodal system accessible via every input modality (Caramazza, Hillis, Rapp, & Romani, 1990; Fodor, 1983) or, on the contrary, as a multimodal system with separate verbal and visual semantic stores (Beauvois, 1982; McCarthy & Warrington, 1988)? Although the jury is still out on this last point (Gainotti, 2011, 2012), semantic storage deterioration is usually characterized by crossmodal disorders, that is, it manifests itself across different formats of stimulus presentation and response modalities (Warrington & Shallice, 1979). Considerable progress has been made in our understanding of semantic memory, through observations of patients presenting with category-specific semantic deficits (see case review by Capitani, Laiacona, Mahon, and Caramazza (2003)). These case studies have inspired new cognitive neuropsychological accounts of semantic system organization, which can be divided into two sets. The first set considers that semantic memory is composed of multiple subsystems that are either partially or totally independent at the functional and anatomical levels. For example, according to sensory/functional theory (SFT) and its variants (Warrington & McCarthy, 1987; Warrington & Shallice, 1984), semantic knowledge about concepts is topographically organized according to the properties that are mostly distinctive for a given category or more relevant during their acquisition. Similarly, according to the domain-specific knowledge (DSK) systems hypothesis (Caramazza & Mahon, 2003; Caramazza & Shelton, 1998), the semantic system is divided into topographically organized domains of knowledge: animals, fruit/vegetables, conspecifics and, possibly, tools. The second set of accounts regards this semantic system as a unitary one, without any explicit functional or anatomical organization. Here, all the features of the different domains of knowledge are brought together within the same distributed network. The internal structure of knowledge is governed by the frequency of co-occurrence between features and the distinctiveness of those features for a given entity. In these models (computational account; Devlin, Gonnerman, Andersen, & Seidenberg, 1998; Gonnerman, Andersen, Devlin, Kempler, & Seidenberg, 1997; conceptual structure account; Tyler & Moss, 2001), concepts are therefore represented by shared or

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distinctive features. This theoretical framework was essentially derived from studies of patients with Alzheimer's disease or semantic dementia, and was particularly inspired by the time course of these patients' gradual loss of conceptual knowledge. Critically, those two sets of models make very different predictions. Multiple subsystems accounts assume that one subsystem breakdown will result in a category-specific deficit. By contrast, in unitary accounts, internal structure of knowledge relies upon a differential probabilistic vulnerability between features. Any category-specific deficit will therefore result from impairment of features assumed to be the most vulnerable to pathology.

Semantic dementia (SD) is frequently regarded as a model of progressive semantic breakdown. It is a type of lobar degeneration characterized by the gradual loss of conceptual knowledge (Moreaud et al., 2008; Neary et al., 1998; Snowden, Goulding, & Neary, 1989; Warrington, 1975), resulting in limited vocabulary in speech (anomia), poor comprehension and deficits in the identification not just of objects and persons, in both the visual and verbal input modalities, but also of smells, tastes and sounds (Bozeat, Lambon Ralph, Patterson, Garrard, & Hodges, 2000; Luzzi et al., 2007; Snowden, Thompson, & Neary, 2004). This selective impairment in semantic memory occurs without any generalized intellectual impairment, deficit in day-to-day memory or visual perceptual abilities. Moreover, language remains fluent, free from syntactic errors, well-structured and without any phonological deficits. This syndrome arises out of temporal lobe atrophy, often bilateral but predominantly on the left side (Hodges, Patterson, Oxbury, & Funnell, 1992). Atrophy is particularly pronounced in the inferolateral areas of the anterior temporal lobes, now known to be a core region supporting semantic cognition (Binney, Embleton, Jefferies, Parker, & Ralph, 2010; Lambon Ralph, Pobric, & Jefferies, 2009; Patterson, Nestor, & Rogers, 2007; Pobric, Jefferies, & Ralph, 2010; Rogers et al., 2006; Visser, Embleton, Jefferies, Parker, & Ralph, 2010; Visser, Jefferies, & Lambon Ralph, 2010).

Category-specific deficits have rarely been reported in SD cases. However, tasks involving naming to description, description-to-picture matching and verbal definitions yielded a disproportionate breakdown for sensory/perceptual features compared with functional/associative features (Lambon Ralph, Patterson, Garrard, & Hodges, 2003). Nevertheless, four SD cases with a category-specific semantic deficit have been documented in the literature. In only one of these cases was there evidence of better preservation for living categories (Patient IW; Lambon Ralph, Howard, Nightingale, & Ellis, 1998), with relatively poor performance on perceptual attributes. By contrast, the other three SD cases (MF; Barbarotto, Capitani, Spinnler, & Trivelli, 1995; LI; Zannino et al., 2006, and KH; Lambon Ralph et al., 2003) exhibited the reverse pattern of deficits, with poorer performance for living things. While perceptual features were more impaired than functional ones for KH and LI, no significant difference was found between the two for MF. Temporal lobe atrophy was bilateral, but more pronounced on the right side for MF and KH, whereas LI's temporal lobe atrophy was more pronounced on the left side, with atypical diffuse atrophy extending to the parietal regions. By contrast, IW's temporal lobe atrophy was left-sided only, with an intact right temporal lobe.

It is important to emphasize that both Barbarotto et al. (1995) and Lambon Ralph et al. (2003) described a domain-specific deficit, focusing their analysis on living versus nonliving entities rather than a discrete category-specific one (i.e., between animals versus vegetables knowledge for example).

We could highlight another critical point from those previous studies. Given that SD patients typically present with expressive and receptive language complaints (Belliard et al., 2007), it may be unwise to assess semantic knowledge with tasks that rely on verbal expressive behavior. Despite this, of the five tasks making up Lambon Ralph et al.'s (2003) semantic assessment, three necessitated verbal output. Similarly, their assessment of semantic attribute knowledge (i.e., functional or sensory) required participants to understand short sentences describing an item, then either to name or point to the item in question. Therefore, in our opinion, the possibility of a category effect on the SD patients' performance cannot be ruled out.

We are not aware of any previous group study that has specifically investigated the effect of category on semantic knowledge in SD, despite the fact that contemporary cognitive accounts of semantic knowledge organization are largely based on category-specific effects.

The aim of this study was thus to determine whether a category-specific semantic effect could be highlighted by means of a semantic sorting task administered to a large cohort of SD patients.

2. Methods

2.1. Participants

2.1.1. Semantic dementia

Between 1991 and 2007, 55 patients who fulfilled the diagnostic criteria for SD (Neary et al., 1998) were followed up at the memory clinic of Rennes University Hospital (Belliard, Merck, Jonin, Lemoal, & Vercelletto, 2011). All of them presented with the typical clinical features of SD: a history of complaints about worsening comprehension deficits, anomia, and difficulty identifying objects and/or persons, reflecting a predominant and distressing loss of conceptual knowledge, contrasting with the relative preservation of day-to-day memory and perceptual abilities. Speech was still fluent, without any phonological or syntactic errors. Of these 55 patients, 35 were administered a complete neuropsychological battery within 3 months of diagnosis (see Table 1), together with a 64-item semantic sorting task. The results of the background neuropsychological assessment are summarized in Table 2.

Many of them (thirteen patients) performed below normal levels on the Mini Mental State Examination (MMSE; Folstein, Folstein, & McHugh, 1975), essentially due to comprehension

Table 1
Demographic and clinical features.

	Semantic dementia ($n = 35$)		Alzheimer's disease ($n = 10$)		Controls ($n = 12$)	
	Mean (SD)	Range	Mean (SD)	Range	Mean (SD)	Range
Age	62.7 (6.4)	48–73	74 (7.9)	59–87	62.8 (4.5)	55–70
Sex (M:F)	25:10		3:7		5:7	
Education (in years)	9.5 (3)	7–18	10.7 (3.7)	7–20	11 (4.7)	7–17
Illness duration (in months)	36 (19.6)	12–96	30.7 (13.6)	12–60		
Side of atrophy (Left:bilateral:right)	15:17:3					
Severity of atrophy						
Left side	2.1 (0.65)	0.5–2				
Right side	1.36 (1.07)	0–1				