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# Loss of Semantic Memory: Implications for the Modularity of Mind

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We report a patient, PP, with semantic dementia who was studied longitudinally over two years. During this period she showed a progressive and profound loss of semantic memory affecting factual knowledge, vocabulary, and object knowledge via all sensory modalities. In the face of this near total dissolution of semantic memory, we have addressed the issue of the fate of other cognitive processes. Our findings suggest that nonverbal problem solving, auditory verbal and spatial short-term memory, the high-level visuoperceptual abilities involved in object constancy, and some basic syntactic processes may operate independently of semantic memory and are therefore independent cognitive modules. In contrast, the integrity of both the phonological representations of words used to produce speech and the representations (or structural descriptions) used to recognise familiar objects appear ultimately to depend on semantic memory.

## INTRODUCTION

One of the major goals of cognitive neuropsychology is to understand how knowledge about the world is represented and organised. The study of brain-damaged patients with disorders of knowledge (or semantic memory)

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has already contributed significantly in this regard. Particular attention has been devoted to issues concerning the internal architecture of semantic memory, for instance, whether there is separate representation of knowledge relating to different sensory modalities, or to different conceptual categories, or both (e.g. Hart, Berndt, & Caramazza, 1985; Hillis & Caramazza, 1991; McCarthy & Warrington, 1990; Pietrini et al., 1988; Sacchett & Humphreys, 1992; Sheridan & Humphreys, 1993; Warrington, 1975; Warrington & Shallice, 1984). A question that has received less intense scrutiny is the relationship between semantic memory and the input/output systems that, in normal processing, are in constant interaction with semantic knowledge (see Schwartz & Chawluk, 1990, for one of the very few discussions of this issue). Can these putatively separate modules be maintained intact when semantic memory deteriorates? We have had the opportunity to address this question in a unique patient with a progressive and profound loss of semantic knowledge. It should be noted that we use the term "module" merely to denote a cognitive process or subsystem that is considered functionally separate from other subsystems in contemporary information processing models, rather than in the strict sense as defined by Fodor (1983; see also Moscovitch & Umilta, 1990). Our usage of module and modular concurs, therefore, with Shallice (1988).

Semantic memory is the long-term memory system that represents knowledge of objects and their inter-relationships, facts and concepts, as well as words and their meaning (Kintsch, 1980; Tulving, 1972; 1983). In contrast to episodic or event memory, semantic memory is culturally shared, not temporally specific, and, to a large extent, acquired early in life.

Patients with dementia of Alzheimer type (DAT) are characteristically impaired on tests of semantic memory, and most authors interpret these impairments as a breakdown in the structure of semantic memory as the disease progresses (Bayles & Tomoeda, 1983; Huff, Corkin, & Growden, 1986; Hodges, Salmon, & Butters, 1990; 1991; 1992b; Martin & Fedio, 1983). In DAT, however, there is virtually always accompanying severe impairment of episodic memory and, eventually, disruption of many other cognitive domains as well, including visuo-spatial and frontal "executive" function. This heterogeneity of deficits is an obstacle to the investigation of at least some issues regarding semantic memory. To understand the inter-relationship of semantic memory and other cognitive processes, it is necessary to study patients with more selective deficits of semantic memory.

Selective impairment of semantic memory was first clearly described by Warrington (1975), who reported three patients with cerebral atrophy presenting with progressive anomia and impaired word comprehension. Detailed neuropsychological testing revealed a loss of receptive and expressive vocabulary, and impoverished knowledge of a wide range of living things and inanimate objects; specific subordinate (attributional) knowledge was particularly affected. Warrington, drawing on a distinction made by Tulving (1972), identified the deficit as one of semantic memory. Since this seminal paper, a number of other patients with selective semantic memory impairment have been reported. The majority have occurred in the context of extensive neocortical damage, for instance following herpes simplex virus encephalitis (e.g. De Renzi, Liotti, & Nichelli, 1987; Pietrini et al., 1988; Sartori & Job, 1988; Sheridan & Humphreys, 1993; Warrington & Shallice, 1984). As in the case of DAT, many of these patients have also had additional complicating cognitive deficits, making them less than ideal candidates for addressing the question of the independence of semantic memory from other putative cognitive modules.

Some patients with relatively purer semantic memory impairment have been reported under the label of "primary progressive aphasia." The latter term was introduced by Mesulam (1982) to describe a syndrome characterised by a progressive disorder of language with sparing of general cognitive abilities. Since Mesulam's description, however, this term has been applied to patients with linguistically very different disorders. The majority of patients, like the original Mesulam cases, have presented with a progressive non-fluent aphasia with prominent impairment of the phonological and syntactic aspects of language, but relatively preserved comprehension. Other cases described under this rubric, by contrast, have had clear semantic memory loss causing deficits in both word production and comprehension, but with relative sparing of other components of language (e.g. Basso, Capitani, & Laiacona, 1988; Poeck & Luzzatti, 1988; Tyrrell, Warrington, Frackowiak, & Rossor, 1990). To avoid confusion, and to encapsulate the key component of the disorder, we have adopted the term "semantic dementia" (Hodges, Patterson, Oxbury, & Funnell, 1992a; Patterson & Hodges, 1992; Saffran & Schwartz, in press; Snowden, Goulding, & Neary, 1989) to describe a form of progressive fluent aphasia with the following characteristics: (1) selective impairment of semantic memory causing severe anomia, impaired single-word comprehension (both spoken and written), reduced generation of exemplars on category fluency tests, and an impoverished fund of general knowledge; (2) relative sparing of other components of language output, notably syntax and phonology; (3) normal perceptual skills and non-verbal problem solving abilities; (4) relatively preserved autobiographical and day-to-day (episodic) memory; and (5) a reading disorder with the pattern of surface dyslexia.

This report describes our longitudinal investigation of one patient with semantic dementia, PP, who first presented to us in 1990 with an apparently pure loss of semantic knowledge. Although this deficit was already severe at initial assessment, it has shown a further dramatic progression over two subsequent years, revealing some important facts about the organisation of semantic memory. More significantly, we believe that we have demonstrated a crucial role of semantic memory in several processes usually considered to be independent of semantic memory: the integrity of the speech output (phonological) lexicon, the ability to read aloud words with an atypical spelling-sound relationship, and the integrity of the stored structural descriptions involved in identifying objects may all depend on intact semantic memory. Other aspects of cognition that appear to be truly autonomous from central semantic memory include auditory-verbal and spatial short-term memory, non-verbal problem solving, visuo-spatial abilities, the high-level visuo-perceptual processes involved in object constancy, and at least some aspects of syntactic processing.

## CASE REPORT

PP, a 68-year-old, right-handed ex-clerical officer and secretary, presented to us in August 1990 with a 2-year history of progressive loss of memory for names of people, places, and things, together with impaired comprehension of nominal terms. She also complained from the onset of problems in recognising even very familiar people from sight, voice, or description. Her fund of general knowledge was radically impoverished. When asked, Have you ever been to America? she replied "what's America?", or asked, What's your favourite food?, she replied "food, food, I wish I knew what that was." Despite this profound deficit in semantic memory, her day-to-day memory remained fairly good. She could remember appointments and keep track of family events. Sadly, she retained insight into her deficit and at times became severely depressed. There had been no deterioration in self-care and at that time she was able to groom, dress, and feed herself without any difficulty.

Spontaneous speech was well articulated with normal prosody. She was able to produce fluent and grammatically correct sentences, but conversation was punctuated by severe word-finding difficulty and frequent semantic paraphasias (an example of her speech output is given later). Phonemic errors were never observed. The degree of anomia was such that she never correctly named a single item (whether presented as picture or real object), either on informal testing or on a number of formal naming tests (see following). Her responses were usually "don't knows" or vague descriptions with occasionally some indication of broad superordinate knowledge, such as "is it an animal?". PP responded appropriately to simple questions about personal events (e.g. "how are you today?"; "is your mother well?"), but there was a profound deficit in understanding of all but very common and rather general nominal terms (such as mother, today, home, etc.), with the result that, from the earliest assessment, she was at or near chance levels on word–picture matching tests. On a wide range of word fluency tasks, she was unable to generate exemplars from any category (e.g. animals, fruit, vegetables, musical instruments, vehicles, household items, etc.). Repetition of single words was normal, and she was even able to repeat grammatically complex sentences (with a restricted vocabulary—the sentences from the TROG; Bishop, 1983). Reading was severely disrupted, with a letter identification deficit and a tendency to read letter-by-letter; but if a word with regular spelling-to-sound correspondence was spelled aloud to her, she could translate the letter string into a correct pronunciation. Similarly, she managed to write to dictation some regular words. She was able to do simple mental arithmetic, indicating that she still knew the "meanings" of numbers. In contrast to her profound language impairment, her visuo-spatial abilities were remarkably well preserved; copies of complex geometric shapes were executed flawlessly (see Hodges et al., 1992a).

CT and MRI scan revealed a moderate degree of cerebral atrophy, more marked in the left hemisphere, particularly around the sylvian fissure. A PET study revealed hypometabolism confined to the temporal and adjacent parietal regions of the left hemisphere (for details see Patterson, Graham, & Hodges, 1994).

On follow-up over the subsequent 12 months (August 1990 to July 1991), there was only moderate change in her general abilities. She remained self-caring and was able to do very simple housework, but when shopping she no longer knew which items to choose, and similarly her cooking ability deteriorated because of a lack of understanding of the fundamental processes and utensils involved. She enjoyed seeing family members and her visits to Cambridge. Over the next year (August 1991 to September 1992), however, there was a marked decline in her everyday abilities. She has become unable to dress appropriately, has stopped doing all housework and uses common items (such as a hairbrush and spoon) inappropriately. Even close family members are no longer recognised.

Her language abilities have deteriorated further. Her spontaneous speech now contains virtually no nouns and few verbs other than general ones like "come" and "do," and she tends to produce grammatically correct and fluent stereotyped phrases such as "Oh dear, I wish I could think what I wanted to say." Indeed, on most recent testing, virtually her only spontaneous utterance has been the sadly appropriate phrase "I don't understand at all." It is now extremely difficult to get her to comply with any tests, although, as we will describe later, her auditory verbal short-term memory remains intact and her visuo-spatial skills are still reasonably retained.

## STANDARD PSYCHOMETRIC TEST DATA

The results of standard clinical neuropsychological tests performed in August 1990 are shown in Table 1. As in the other patients with semantic dementia (Hodges et al., 1992a), there was a discrepancy between verbal and performance IQ in favour of the latter, with a characteristic subtest profile. Age-scaled scores on the Vocabulary, Information and Similarities subtests were severely impaired, in contrast to the exceptionally good score for Digit Span. Of the Performance subtests, Block Design and Digit Symbol were relatively well preserved. PP's failure on the Picture Completion subtest probably reflects a failure to comprehend the stimuli, as evidenced by her performance on an object decision test (see following). The Object Assembly and Picture Arrangement subtests were not performed. Her performance on the Raven's Coloured Progressive Matrices (Raven, 1962) was almost perfect. She copied the Rey Complex Figure

TABLE 1
Basic Neuropsychological Data When First Tested in 1990 (Maximum Scores are Shown in Parentheses)

| | *PP* |
|---|---|
| WAIS Subtest Scores (age scaled) | |
| Information | 1 |
| Digit Span | 17 |
| Vocabulary | 4 |
| Arithmetic | 7 |
| Similarities | 0 |
| Block Design | 12 |
| Digit Symbol | 8 |
| Picture Completion | 4 |
| Verbal IQ (prorated) | 74 |
| Performance IQ (prorated) | 97 |
| Rey Figure Copy (36) | 34 |
| 45min delayed recall | 8 |
| Raven's Coloured Matrices (36) | 35 |
| Facial Recognition Test (54) | 42 |
| Judgement of Line Orientation (30) | 25 |
| Digit Span | |
| Forward | 9 |
| Backward | 6 |
| Logical Memory WMS[a] | |
| Immediate (21) | 0 |
| 30min delayed | 0 |

[a]WMS = Weschler Memory Scale.

without any major error and scored within the normal range for her age on the Benton Judgement of Line Orientation and Facial Recognition Tests (Benton, deS Hamsher, Varney, & Spreen, 1983).

## EXPERIMENTAL TESTS OF SEMANTIC KNOWLEDGE

### Semantic Memory Test Battery of Hodges et al. (1992b)

To explore PP's declarative semantic knowledge, in August 1990 we administered a standardised battery of tasks designed to assess various types of input to, and output from, central representational knowledge about the same set of items. This battery has been described in detail previously (Hodges et al., 1992a; 1992b). It contains 48 items (from the corpus of line drawings by Snodgrass and Vanderwart, 1980) chosen to represent 3 categories of animals (land animals, sea creatures, and birds) and 3 categories of man-made items (household items, vehicles, and musical instruments), with the items across categories matched for prototypicality. In brief, the five sub-tests consist of:

1. Category fluency for each of the six main categories plus two lower-order categories (breeds of dog and types of boat).
2. Naming of all 48 line drawings.
3. Picture sorting at three levels; superordinate (living vs. man-made), category (land animal vs. water creature vs. bird), and subordinate/attributional (e.g. British vs. foreign animal, electrical vs. non-electrical household item).
4. Identifying one of six pictures (in within-category arrays) in response to a spoken object name (e.g. "point to the trumpet" in a set of six musical instruments).
5. Generation of verbal definitions in response to the spoken name of the item (i.e. how would you describe to someone who has never seen it before—a telephone, fish, helicopter, swan, etc.).

In Table 2, PP's profoundly impaired performance across the range of tests is contrasted with the mean scores from a group of 25 normal subjects matched for age and education (mean age 69.0 ± 8.4, mean years of education 10.7 ± 2.2). PP failed to produce a single appropriate response on any of the three tasks requiring language output: category fluency, object naming, and word definitions. Category fluency was further tested on a wider range of categories (e.g. vegetables, fruit, body parts, clothes, occupations etc.); on none of these was she able to produce a single

TABLE 2
Performance of PP and Normal Controls on a Battery of Tests Designed to Probe Semantic Knowledge Via Different Modalities of Input and Output

| | *PP* | *Normal Controls (n = 25)* |
|---|---|---|
| | *Number* | *Mean (SD)* |
| Category Fluency | | |
| Living (4 categories) | 0 | 46.8 (12.6) |
| Man-made (4 categories) | 0 | 47.7 (14.3) |
| | *% Correct* | *Mean % (SD)* |
| Naming to Confrontation | 0 | 96.7 (2.2) |
| Generation of Definition | 0 | 97.9 (1.5) |
| Word-to-picture Matching[a] | 27 | 99.8 (0.1) |
| Sorting of Pictures | | |
| Level I: Living vs. man-made[b] | 100 | 100.0 (0.1) |
| Level II: Superordinate category[c] | 72 | 95.6 (2.5) |
| Level III: Subordinate attribute[b] | 42 | 96.5 (3.9) |

[a]Chance level 17%. [b]Chance level 50%. [c]Chance level 33%.

exemplar and she usually replied with a statement such as "I wish I could remember what a fruit was." The only task requiring verbal output on which PP managed to generate some appropriate responses was a fluency test using initial letter as the retrieval cue. Her total for the letters F, A, and S was 13 (controls' mean total = 44.5 ± 10). Not surprisingly, her responses were high-frequency words of the sort that also appeared in her restricted spontaneous conversation (e.g. fair, feel, after, silly, sunny, sister, etc.).

Matching spoken names to pictures from within-category arrays was not significantly above chance, nor was sorting of pictures according to attributes (42% in a 2-choice task). In keeping with prior observations on patients with semantic memory impairment (e.g. Warrington, 1975), however, PP's ability to sort pictures into superordinate (i.e. living and nonliving) categories was initially perfect (48/48). Sorting the same pictures at a category level (e.g. musical instrument vs. vehicle) was significantly impaired but above chance (72% with 2 choices). The results of longitudinal assessment of her picture sorting ability are shown in Fig. 1. Performance at the superordinate level remained perfect for the first year of follow-up, but then fell progressively over the subsequent year to 47% (chance = 50%) on the latest assessment. Sorting at the category level fell rapidly to chance in the first few months of follow-up.

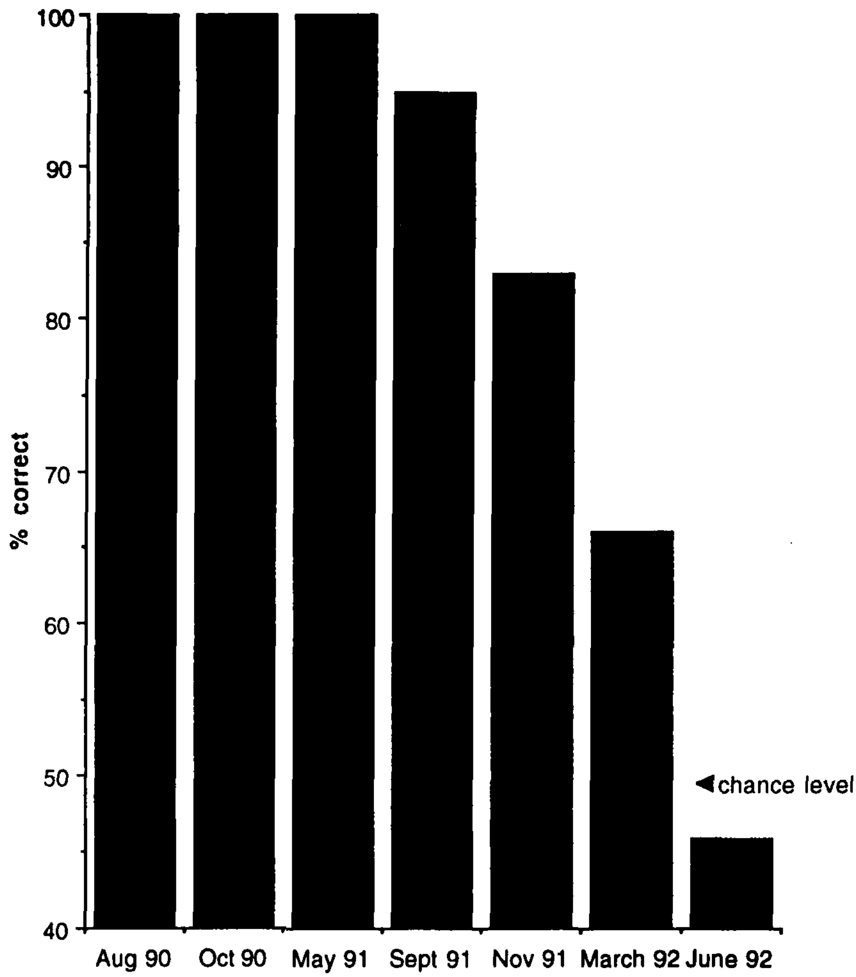



FIG. 1 PP's performance over time at sorting pictures into living and man-made from the Semantic Memory Test Battery of Hodges et al. (1992b).

## Picture–word Matching with Semantic and Visual Distractors

In October 1990 we gave PP the auditory version of the word–picture matching test from the PALPA battery (Kay, Lesser, & Coltheart, 1992). In this, the subject is presented with an array of five pictures consisting of the target, a closely related semantic foil, a distantly related semantic foil, a visually related foil and an unrelated foil from the same category as the visual foil. Normal controls achieve a mean score of 38.8/40 (97%) ± 2.2.

PP scored 14/40 (35%), barely above the chance level of 20%. Her errors did, however, show a hint of retained knowledge of the semantic field of the target item: there was 1 refusal to respond and, of the remaining 25 errors, 10 were close semantic, 6 distant semantic, 5 visual, and 4 unrelated. (See Funnell, 1992, for discussion of performance on this test in the context of progressive loss of semantic knowledge.)

## Tests Involving Real Objects and Toy Animals

We also attempted to assess PP's knowledge of real objects. In February 1991 she was presented with 25 everyday household items from one of the authors' homes, and was asked "what would you do with this?" Her responses were scored on a 0 to 2 scale. A score of 2 was given when she indicated by any means (spoken or gestural) the correct and specific use of the object (e.g. napkin, "we put it on there [gesturing to her lap] when we eat"; hairbrush, "you do this with it [pointing to her hair], you've got to keep it down"). A score of 1 was awarded when she indicated correct category membership only (e.g. spoon, "for eating"; letter opener, "you cut something"), and 0 was given for completely incorrect or non-responses. Of her 25 responses, 8 received scores of 2 (hairbrush, toothbrush, napkin, comb, envelope, ladle, and bowl); 7 were given scores of 1 (fork, pepper grinder, spoon, knife, letter opener, whisk, and grater), and the remaining 10 responses scored 0 (e.g. sewing cotton, "is it something we eat?"; salt shaker, "something like water goes in there"; tin opener, "I don't remember seeing one of these"; eraser [after smelling it], "don't know"; toothpaste "do you put it on here?" [pointing to her fingers]; candle, needle, box of matches, and tin of tuna, all "don't knows"). She was then given the same items in groups of four or five and was asked to arrange them in pairs or triplets according to their common usage (examples of intended groupings were candle and matches; toothbrush and toothpaste; needle, thimble, and sewing cotton, etc.). She produced none of the intended groupings with the exception of the cutlery (knife, fork, spoon). She paired the comb with the toothpaste, and the tin of tuna with the ladle.

On another occasion, early in 1991, we attempted to test her ability to discriminate between different animals using 20 plastic models. She was unable to understand the meaning of terms such as farm, zoo, fierce, etc.; on the other hand, she did seem to understand, or at least responded to, questions about whether an animal was one that she might see near her home, and whether it would be large or small (all of the model animals were of roughly equivalent size); however, her performance on both of these discriminations was at a chance level.

## Environmental Sound–Picture Matching

In order to test PP's knowledge of environmental sounds, in November 1990 we administered a test in which the subject is presented with examples of everyday sounds and is required to select, from an array of six pictures, the one that goes with each sound. A pre-recorded tape was used and PP was allowed repeats of the sound recording if requested. Each of five arrays consisted of items from the same category, and the categories were: sounds made by a person (e.g. crying, laughing, sneezing); everyday activities (e.g. brushing teeth, eating, drinking); household items (e.g. clock, telephone, vacuum cleaner, etc.); transport (e.g. car, motorbike, helicopter); and animals (e.g. cat, dog, cow, sheep). Controls were not administered the test but should perform perfectly. Since all of the items in a single array were tested consecutively, the appropriate chance level of this test is 37%; PP achieved a score of 13/30 (43%). Occasionally she responded not by pointing to one of the picture alternatives but with her own interpretation of the sound; for instance, in response to the recording of a motorbike, she replied "it sounds like water," and after listening to the recording of human crying, she said "it sounds like rain." In most cases she simply said "no idea" and pointed at random to one of the pictures.

## Famous Face and Name Identification

Since one of the features noted by PP's family at the time of presentation was her difficulty in identifying familiar people, we attempted to explore PP's knowledge of famous personalities, using initially an updated version of the Famous Faces Test. This test consists of 50 faces of personalities who were famous for a relatively limited period between the 1940s and 1980s, with 10 items per decade (see Hodges & Ward, 1989, for further detail). Normal subjects name approximately 60% of the faces from each decade and are able to identify most of the others by description (e.g. "it's that ex-film star who was the US president"). PP was unable to name or identify by description any of these faces. We therefore selected a different group of 10 extremely famous faces (e.g. Winston Churchill, Charlie Chaplin, John F. Kennedy, the Queen, Margaret Thatcher). PP was unable to name or describe any of these. Furthermore, she did not recognise them as familiar or famous, and was unable to select the correct name to go with the face when presented with three alternatives (e.g. is this Fred Astaire, John F. Kennedy, or Harold Wilson?). She reported that none of these names were familiar to her. We then made up arrays consisting of one extremely famous face with two unknown faces. Her ability to point to the familiar face in each array was no better than chance. In addition, we informally probed her knowledge of these very famous personalities. One example is given next.

Examiner: One of these people is a member of the Royal Family, do you know which one?
PP: No.
E: Which one is the Queen?
PP: I've heard of the Queen, but I wish I knew what it was.
E: Is the Queen a man or a woman?
PP: (long pause) . . . I think it's a woman but I'm not sure.

To test her face-matching ability, we constructed six ad hoc arrays of six photographs, each array containing two non-identical photographs of the same famous person. PP scored perfectly when asked to indicate which two were the same person. This preserved perceptual ability was in keeping with her performance on the Benton Facial Recognition Test (Benton et al., 1983) and other visually based tasks (see earlier) that do not require semantic knowledge of the items presented.

## Comment

Assessment of PP's semantic memory revealed a profound deficit in all categories, independent of the modality of access or output. At least on these "declarative" tests of semantic knowledge (see later for comments on implicit measures of semantic memory), we were unable to demonstrate any islands of preserved knowledge, except that—for the first year of our longitudinal study—her classification of pictures as living or man-made items was flawless. This finding of initially perfect superordinate knowledge, impaired (but above chance) category level knowledge and a complete absence of subordinate knowledge addresses issues about the organisation of semantic memory, which will be discussed further later. PP's deficit in semantic memory was apparent not only with pictures and words as stimuli but also with real objects, model animals, and environmental sounds. In addition, she showed a profound agnosia and loss of knowledge for very famous people, with preserved ability to match faces (familiar or unfamiliar) by identity.

# ASPECTS OF COGNITIVE ABILITY APPARENTLY INDEPENDENT OF SEMANTIC MEMORY

In this section we describe our investigation of cognitive abilities which, on the basis of PP's performance, appear to be relatively independent of semantic memory. In other words, at a time when PP demonstrated profound semantic impairment, her performance in these areas was either within normal limits or at least showed a striking degree of preservation.

## Syntactic Abilities

### *Syntactic Aspects of Speech Production*

PP's spoken language, though empty and somewhat disrupted by word-finding difficulty, has always been syntactically well formed, as illustrated by the following transcript of a conversation with the authors in the autumn of 1990.

PP: My mother is 98, and my mother-in-law is 95 [*note: both of these facts were correct*] . . . that's, that's, so we have to go down there in er . . . quite often, near my mother . . . near my daughter, I mean.
Examiner: Are they well?
PP: They're all . . . they're not too bad, both of them. My mother is 98, she lives with my . . . sister. And we've been staying with them tonight, we've been down south to see the little girl [*note: she is referring to her grand-daughter*].
E: What are your main problems at the moment?
PP: Main problems . . . thinking about . . . thinking about . . . I've been worried to death thinking, trying, I'm going to try and think with you today.
E: That worries you quite a bit, doesn't it?
PP: Yes, because I . . . I think of everything but I . . . I can't always talk.
E: Is the problem with thinking or talking?
PP: I don't know . . . I don't . . . I can't, I think of things . . . I can't often say . . . er . . . say what to say.
E: How is your memory?
PP: It's very bad.
E: Do you remember seeing us before?
PP: Yes, oh yes, I *do*. I can remember *that* sort of thing.
E: Have you been in this room before?
PP: Yes, we came here before [*note: correct*].

Two years later, her language comprehension and production had become extremely limited, with stereotyped and even rather echolalic responses, as shown by the following conversation in July 1992:

E: Where do you live?
PP: Where do you live . . . I don't understand at all.
E: Where's your house?
PP: House . . . I don't understand at all.
E: That's a nice dress you've got on today.
PP: Yes it is.
E: Did you choose it?
PP: Choose it . . . I don't understand at all.
E: Have you seen your mother recently?
PP: Yes.
E: How old is your mother now?
PP: I don't understand at all.

### *Explicit Syntactic Comprehension Tasks*

Provided that the vocabulary was tailored to her very restricted knowledge of referential meaning, PP's performance on two standard measures of syntactic comprehension was reasonably competent when tested in August 1990. First of all, on a simplified version of the Token Test (De Renzi & Faglioni, 1978), with tokens varying on only 2 dimensions (black and white, large and small circles) she was able to obey 10/12 complex commands. She responded correctly to the following structures: next to, on top of, between, touch X with Y, touch X then Y, don't touch Y touch Z, touch not only Y but also Z. Secondly, PP was administered the Test for the Reception of Grammar (TROG—Bishop, 1983). In this test, the subject is asked to point to the picture (from a set of four) corresponding to a spoken sentence; distractor pictures are designed to discriminate errors arising in syntactic processing from problems with lexical meaning. Each block of four items tests comprehension of the same syntactic structure. The test is graded in difficulty: The initial blocks are easier, and include negatives ("the boy is not running") and plurals ("they are jumping over the wall"); the later blocks include reversible passive sentences ("the girl is being chased by the horse") and sentences with embedded clauses ("the cat the cow chases is black"). Although PP's performance on this test was far from perfect (46/88; 68% correct; normal controls 98 ± 3%), it was significantly better than chance (25%), and almost half of her errors were lexical rather than grammatical.

### *Implicit Syntactic (and Semantic) Processing*

All of the tests described so far require some explicit access to semantic or syntactic knowledge, and thus may underestimate the extent to which patients' knowledge remains intact (Tyler, 1988; 1992). Tasks which tap implicit access do not generally suffer from this problem. In view of PP's exceedingly poor performance on most of the tests already described, we decided in September 1991 to test her ability to use semantic and syntactic information in the process of understanding a spoken sentence by means of an implicit task—the word-monitoring task developed by Marslen-Wilson and Tyler (1980). In this, subjects listen to spoken sentences and are asked to press a response key as soon as they hear a pre-specified target word in the sentence. A patient's response times (RTs) in monitoring for target words in different kinds of sentential context, contrasted with the profile of control subjects' RTs, provides a measure of the normality (or otherwise) of processes involved in the on-line, or immediate, analysis of a spoken utterance.

In this study, PP and control subjects monitored for target words occurring in various word-positions across three different kinds of prose materials: normal sentences, anomalous sentences (which are grammatical but meaningless), and scrambled sentences. In other words, there were two major independent variables: the type of sentence in which the target word appeared (normal, anomalous or scrambled prose) and the position of the target in the sentence (early, middle or late). Each sentence containing a target word was preceded by an initial short sentence providing a minimal context for the interpretation of the target sentence. An example of each of three sentence conditions, with the target word BUS (capitalised) in early position, is:

1. Normal Prose: Bill apologised for being in such a hurry. He said the BUS always left on time and he didn't want to miss it.
2. Anomalous Prose: Fred argued over trying in such a tooth. It said the BUS always tells in space, and he didn't hope to guess it.
3. Scrambled Prose: Such for in apologised hurry a being Bill. The said he BUS and want left always he on didn't it time miss to.

For unimpaired listeners, monitoring RTs get progressively faster from early to middle to late position in the sentence, for both normal and anomalous prose but not for scrambled prose. These word-position effects in normal and anomalous prose reflect the listener's ability to use semantic and syntactic information to construct an interpretative representation of an utterance as it is being heard, or "on line" (Marslen-Wilson & Tyler, 1980). Furthermore, normal subjects show a marked RT advantage for targets in normal prose sentences, relative to anomalous prose, indicating their comprehension and use of semantic information in sensible sentences. If PP is impaired in her use of semantic information but retains some degree of syntactic processing ability, then we would expect her to show a word-position effect in both normal and anomalous prose, but no difference between them. That is, neither the absolute RTs nor the word-position curves in the anomalous and normal prose conditions should differ.

*Methods.* The stimuli were constructed from 54 pairs of normal sentences, each of which contained a high-frequency concrete noun as target word in the second sentence of the pair. For one-third of each of the stimuli, the serial position of the target was early (word-positions 2–4), middle (positions 6–8) and late (positions 10–14) in the sentence. Anomalous prose versions of the normal prose sentences were made by replacing each content word (except the target) with another word of the same form-class and frequency, so that the sentences, although semantically anomalous, were still gramatically structured. Scrambled strings were made by pseudo-randomly mixing the words in normal prose so that the resulting strings had no structure. The position of the target word was unchanged throughout these manipulations. All stimulus strings had been pre-recorded on tape, with a native speaker of British English reading them aloud as naturally as possible (though inevitably with somewhat more natural-sounding prosody for stimuli in the two syntactically structured conditions).

Two groups of control listeners were tested, 10 with and 8 without some degree of the hearing loss that characterises many people of this age (including PP). Their mean age was 69 years (range: 65–73 years), making them appropriate controls for PP. For both controls and PP, the experiment required three testing sessions, such that the same target word (e.g. BUS in the earlier examples) occurred once only in each session. Each session included equal numbers of sentences in the various conditions.

*Results: Control Subjects.* As shown in Fig. 2, control subjects were faster to monitor for targets occurring in Normal Prose than in Anomalous Prose, and also faster in Anomalous Prose than in Scrambled Strings (Min $F'[2, 133] = 70.43$, $P < 0.01$). Subjects also showed a monotonic relationship between speed of monitoring response and position of the target in the sentence, with fastest RTs to targets in late position (Min $F'[2, 74] = 11.38$, $P < 0.01$). However, the word-position effect applied only to Normal and Anomalous Prose conditions (see Fig. 3a). For targets in Scrambled Strings, RTs were not affected by the position of the target word in the sequence (prose × position: $F^1[4, 64] = 21.596$, $P < 0.001$; $F^2[4, 102] = 2.656$, $P < 0.05$; Min $F'[4, 126] = 2.37$, $P > 0.05$). This distinction between the pattern of RTs in Normal and Anomalous Prose compared to Scrambled Strings supports the claim that the word-position effect reflects the representation that listeners construct as the utterance unfolds over time. Since the words in scrambled strings are unstructured, there is no basis for RTs to be facilitated as more of the sequence is heard.

We also calculated an estimate of the magnitude of this facilitation attributable to word-position, by dividing each subject's difference between early and late RTs by his or her own mean RT for early position, for both normal and anomalous prose conditions. For normal prose, RTs to targets occurring in late word-positions were 11–30% (mean = 22%) faster than RTs to early positions. For anomalous prose, the range was 6–22% (mean = 16%).

*Results: PP.* PP had no difficulty performing this task. Her RTs were fast and within the normal range (mean RT = 451msec compared with a control mean of 415msec [range: 336–501msec]), and she rarely missed a

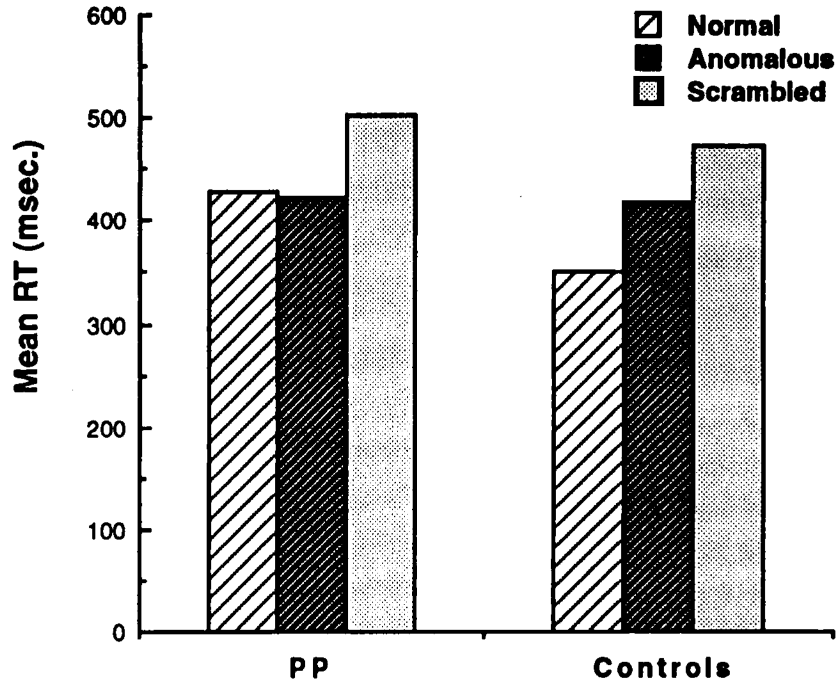
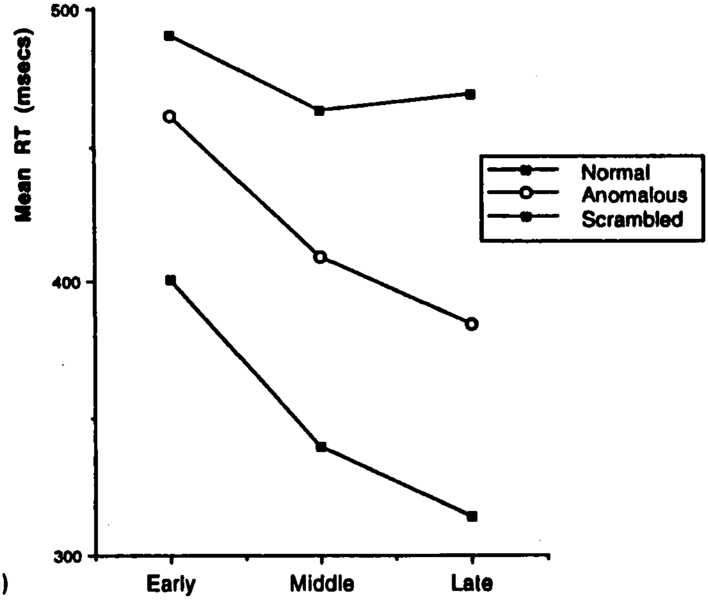



FIG. 2 Prose monitoring experiment: The effects of prose type on mean RT latency compared to that of matched controls.

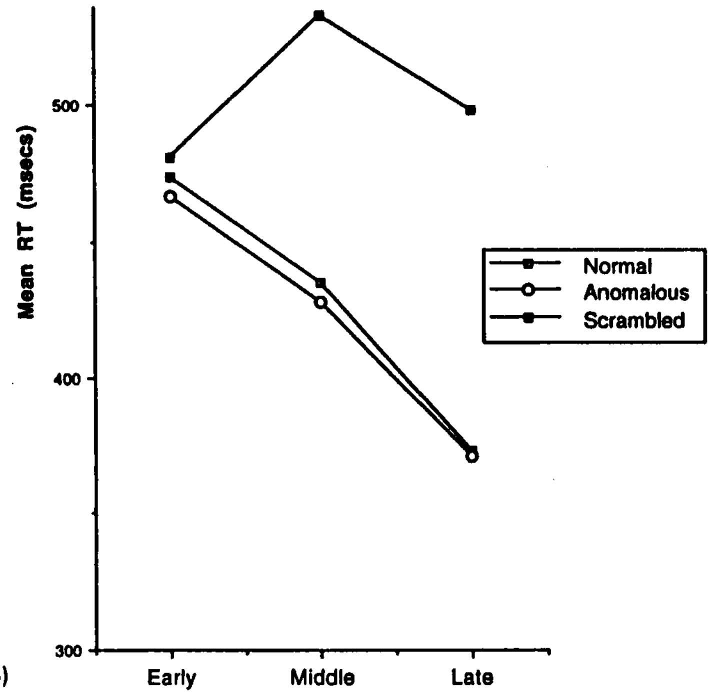
target word. In preparation for the ANOVA, we replaced extreme values (mean ±2 SDs) with the condition mean; only 4% of the data required replacement. PP's mean RTs by prose type are summarised in Fig. 2. A two-factor (prose type × target position) ANOVA was performed on her data. There was a significant effect of prose type ($F[2, 104] = 11.337$, $P < 0.001$). Post-hoc *t*-tests confirmed that this effect was due to faster responses in Normal and Anomalous Prose compared to Scrambled Prose ($P < 0.001$); Normal and Anomalous Prose did not differ from each other ($P > 0.05$).

PP also showed a significant word-position effect ($F[2, 108] = 4.472$, $P < 0.01$). Although the prose by word-position interaction just failed to reach significance ($F[4, 102] = 2.187$, $P = 0.075$), separate ANOVAs carried out on each prose type individually showed a significant word-position effect in Normal Prose ($F[2, 51] = 5.042$, $P < 0.01$) and in Anomalous Prose ($F[2, 51] = 5.37$, $P < 0.01$) but not in Scrambled Prose ($F < 1$). The mean RTs across word-positions in the three types of prose are shown in Fig. 3b.

In all but one respect, PP's pattern of RTs was similar to that of the control group. The single exception is the fact that her RTs were no faster




(a)




(b)

FIG. 3 Prose monitoring experiment: The effects of word position on mean RT latency in controls (Fig. 3a) and patient PP (Fig. 3b).

in Normal Prose than in Anomalous Prose. Figure 3b demonstrates that PP's mean RT at each of the three word-positions was virtually identical in Normal and Anomalous Prose; the difference between the two conditions at each word-position ranged from 1–7msec. In contrast, the means for control subjects were between 60 and 71msec faster for Normal than Anomalous Prose at each word-position.

## Comment

The simplest, most extreme interpretation of the outcome of this experiment is that PP showed essentially normal syntactic processing but a virtual absence of semantic processing. We are, however, only prepared to argue for the weaker conclusion that the data demonstrate differential levels of retained ability in these two domains. The reasons for stopping short of the more dramatic conclusion are as follows.

First of all, with regard to the evidence for normal syntactic processing: Although PP did indeed show an entirely normal word-position effect in both Normal and Anomalous Prose conditions, there may have been a contribution to this effect from the more natural prosodic contours of the sentences in these two conditions than in Scrambled Strings. Furthermore, before concluding that PP had *no* syntactic deficit, one would want to observe adequate performance in other types of on-line tests, such as normal sensitivity to violation of various syntactic constraints (as assessed in experiments by Tyler, 1992; Tyler, Ostrin, Moss, & Cooke, Note 3). We attempted to perform one such experiment that required data from four experimental sessions; sadly, by the last of these test sessions PP's status had declined to the point that—though she might still have been capable of the syntactic demands of the test itself—she could no longer comprehend experimental instructions. Even after extensive examples designed to give her the idea, she responded merely by repeating "I don't understand at all," and we had to abort the test.

Secondly, the predicted and observed absence of any RT advantage for targets in Normal as compared to Anomalous Prose certainly fits with all of our "off-line" evidence of severely disrupted semantic processing; however, (1) the failure to measure a difference between conditions, especially in a single subject, must always be interpreted with some caution; and (2) again, one would want additional evidence from other kinds of implicit semantic processing tasks, for example, semantic priming. In fact, we have carried out a primed word monitoring experiment with PP, where the target in a syntactically unstructured list of words was, on critical trials, preceded by a semantically related word. Although her overall pattern of performance was aberrant from that of control subjects, she did show significant semantic facilitation in some conditions of the experiment (Moss, Tyler, Patterson, & Hodges, Note 2). Thus, once again we hesitate to argue for a *complete* loss of any semantic processing. At least as measured by the paradigm of word monitoring in sentential context, however, PP's performance demonstrates that a substantial degree of syntactic preservation can co-exist with severely disrupted semantic aspects of language. This conclusion is not unique to us: See Breedin and Saffran (Note 1) and Schwartz, Marin, and Saffran (1979) for more extensive data and discussion.

## Non-verbal Problem Solving, Visuo-Spatial, and Object Matching Ability

Longitudinal performance on tasks designed to tap these abilities is shown in Fig. 4. Problem-solving skills were strikingly well preserved in PP: On Raven's Coloured Progressive Matrices (RCPM; Raven, 1962), which consists of progressively more difficult analytical visual problems of a non-verbal type, she continued to score in the high average range for her age during the first 18 months of follow-up, indicating that there was no deterioration in non-verbal intellectual abilities over a period of dramatic decline in semantic memory. Then, between March and June 1992, her performance on the RCPM declined precipitously from 32 to 11.

At presentation in August 1990, PP's performance on copying the Rey Complex Figure was virtually flawless (34 out of a maximum of 36). During the next 18 months, there was only a slight decline (from 34 to 32). Then, over the following 6 months, her performance fell into the impaired range, although she still achieved a score of 8/36 at a stage of floor-level performance on all components of the semantic battery. Similarly, her performance on Benton's Judgement of Line Orientation Test (Benton et al., 1983) was initially well within the normal range (89% correct); and although this declined somewhat over the next 12 months (to 76% correct), once again this level of performance is dramatically superior to that on any semantically based tests. Follow-up after this period was not possible because of her inability to comprehend the test instructions.

Evidence for the dissociation between semantic knowledge and high-level perceptual abilities comes from PP's performance when first assessed in August 1990 using an object matching test (Humphreys & Riddoch, 1984). The subject is presented with an array of three photographs consisting of a target object photographed from a conventional angle, presented above two response alternatives: (1) the same target object photographed from a different (unusual) view, and (2) a photo of a different but visually similar object. The subject is asked to indicate which of the lower two photographs is of the same object as that shown above. The 40 items in the test are divided into those in which the correct alternative represents a fore-shortened view of the object (n = 20) and those classified

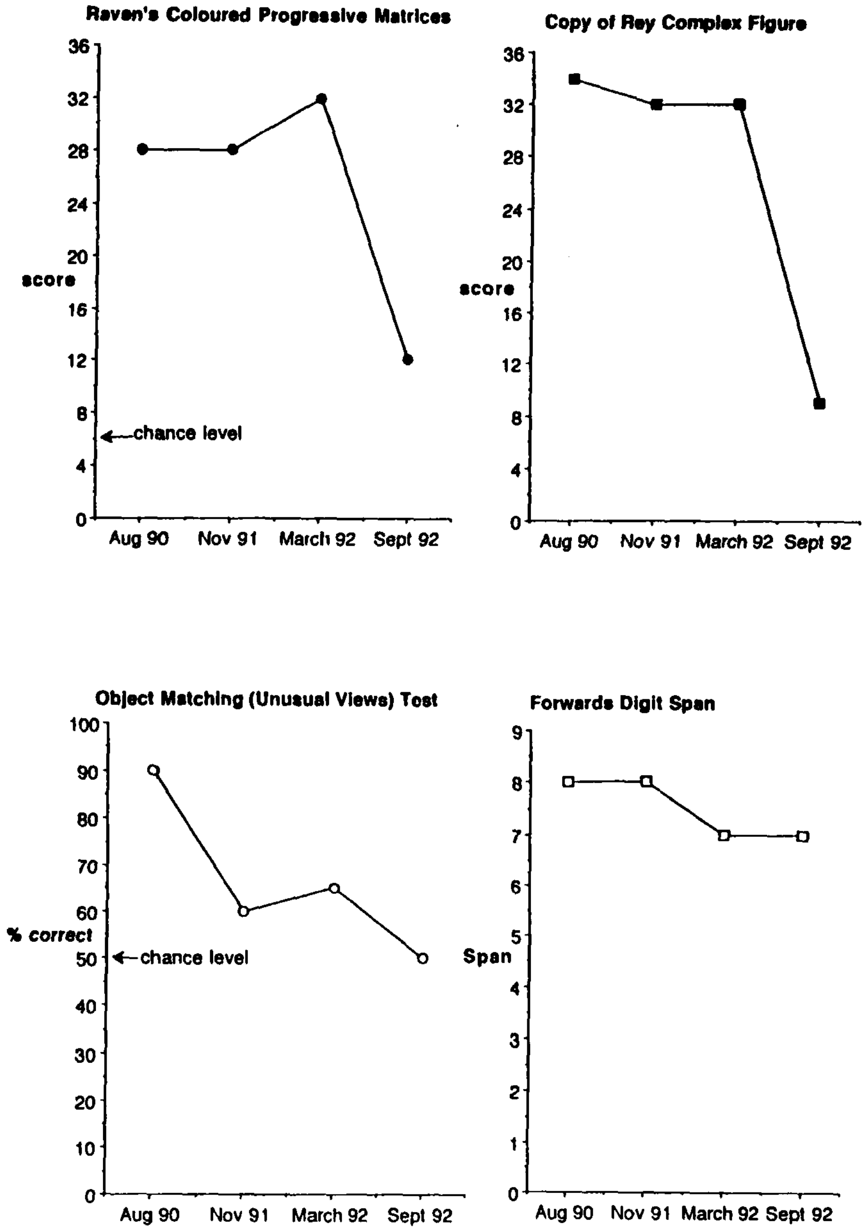



FIG. 4 PP's performance over time on the following tests: Raven's Coloured Progressive Matrices (top left), Copying the Rey Complex Figure (top right), Humphrey and Riddoch's object matching—"unusual views"—test (bottom left), and forward digit span (bottom right).

as minimal feature view (n = 20) (see Humphreys & Riddoch, 1984, for discussion of this distinction). In August 1990 PP performed at a normal level on this task, despite the fact that she could *not* name, identify, or describe the use of any of the objects in the test: PP 19/20 and 18/20 for the foreshortened and minimal feature sets, respectively; controls 18.4 ± 2.0 and 18.7 ± 1.4. This intact object matching, demonstrating high-level visual analytic ability, contrasts sharply with PP's inability to discriminate real from unreal objects (see later). On follow-up over the next 12 months her performance on the object matching test fell rapidly to a chance level.

### Auditory Verbal and Spatial Short-term Memory

Short-term (working) memory also appears to be independent of semantic memory. PP's Digit Span was, on initial assessment, well above normal (nine forward and six backward) and has remained at or near this level over the two years of follow-up (see Fig. 4). Even at the stage when PP indicated essentially no language comprehension and her speech output consisted of simple stereotyped utterances, her forward digit span was eight. On a spatial analogue of the digit span task, the Corsi Block-tapping Test, she achieved a span of six items forward and five backward, which is normal for her age. Longitudinal assessment was not obtained on the Corsi Test.

## IMPAIRMENT IN COGNITIVE ABILITIES DEPENDENT UPON SEMANTIC MEMORY

We turn now to a consideration of those processes which, though theoretically separate modules from central conceptual knowledge, were impaired in PP. It is of course possible that the modules responsible for these processes were independently, structurally damaged; but we will argue that loss of communciation from semantic memory to these other systems is sufficient to account for the observed impairments. We shall focus on two cognitive domains: (1) the phonological representations of words used in speech production and reading aloud, and (2) structural descriptions of objects involved in object recognition.

### Phonological Representations of Words

#### *Repetition*

In contemporary models of language production, the module responsible for the phonological representations of words, the speech output lexicon, is regarded as separate to and hence independent of the semantic system (Butterworth, 1989; Dell & O'Seaghdha, 1992; Levelt, 1992;

MacKay, 1987). According to such a view, whereas a deficit in knowledge of word meaning will of course have devastating consequences for spontaneous speech production, it should not necessarily have any consequences for the ability to repeat spoken words, or to read written words aloud. Indeed, PP was able to repeat single words (regardless of whether she could comprehend their meaning) virtually without error. But as she was also reasonably competent at repeating nonsense words, single-item repetition is insufficient for assessing the status of her phonological representations. Early in our longitudinal study (and rather fortuitously, in the context of a test designed to measure verbal learning of word sequences as a function of repeated presentations), we noticed that PP made a substantial number of errors when she attempted to repeat as few as three of four unrelated words. This seemed a striking phenomenon in a patient with a digit span of nine and unimpaired repetition of sentences (albeit those, from Bishop's [1983] TROG, with a restricted and simple vocabulary of the sort that was still relatively preserved in PP). In order to examine the status of PP's phonological output lexicon, particularly in relation to her semantic knowledge of the words in question, we asked her to repeat lists of "known" and "unknown" words in a version of the task originated by Warrington (1975) and also used by McCarthy and Warrington (1987). PP's "known" words were those that she had recently produced appropriately in spontaneous speech, and also some that she produced on letter fluency tests (see earlier). "Unknown" items were words that we never heard her produce and that she specifically failed to comprehend in picture-pointing tests.

As the results of this experiment have been reported in detail elsewhere (Patterson et al., 1993), we will only summarise them briefly here. There was a striking effect of the known/unknown manipulation both for the three- and four-word lists (see Table 3), with adequate reproduction of lists of words still within PP's vocabulary but very poor performance on

TABLE 3
PP's Repetition of Lists of Known and Unknown Real Words of Length 3 and 4: Proportion of Lists and of Words Correctly Repeated (Tested in May 1991)

| | *Known* | *Unknown* |
|---|---|---|
| List length = 3 | | |
| Lists correct | 0.83 | 0.11 |
| Words correct | 0.94 | 0.66 |
| List length = 4 | | |
| Lists correct | 0.61 | 0.00 |
| Words correct | 0.86 | 0.47 |

lists of previously familiar but now unknown words. This difference was highly reliable by chi-squared tests, whether the measure was number of whole sequences correct or number of words correct. Even more striking, perhaps, than the quantitative performance on "unknown" words was the qualitative nature of PP's errors. She made very few errors of omission, and almost all of her errors of commission involved migrations of phonemes from words within the current sequence being repeated. Her most frequent error, for both monosyllabic and multisyllabic words, was substitution of a different consonant (or consonant cluster) at the onset of the word; a number of these onset errors were perfect "Spoonerisms" such as *mint, rug* → "rint, mug." Treiman and Danis (1988) have demonstrated that normal speakers make frequent errors of this type when asked to repeat sequences of nonsense words, and interpret their results as reflecting the salience of the onset-rime structure of syllables in phonological knowledge.

### Comment

Along with most current models of speech production (see for example Levelt, 1992), we assume that words in the speech-output lexicon are represented as phonological elements rather than as pre-assembled packages. Along with Treiman and Danis (1988), we also assume that the onset-rime syllable division is a prominent aspect of this representation. On the basis of performance by PP (and several other patients with semantic dementia) in repeating known/unknown words, we offer a further hypothesis about the nature of phonological representations. We suggest that the elements of a word's representation are helped to emerge in the right combination not only by virtue of their interconnections at the phonological level, but also by the fact that the whole set of elements making up a word is linked to the word's meaning. This link is activated every time that a person produces a word, and thus could be seen to "reinforce" the integrity of the phonological representation: Just those particular phonological elements, in just that order, respond to activation of that word's meaning. It therefore seems plausible that deterioration of meaning will eliminate an important source of coherence holding phonological elements together in word-based packages. Suppose (to simplify the argument) that the only relevant elements were onset and rime. When PP is asked to repeat a single word like *mint*, there will be only one onset and one rime; and even if there is no meaning to provide binding between these, they can be retrieved and blended for correct output. If a sequence of three or four words is presented for repetition, however, the absence of semantic binding will mean that any active onset may combine with an active rime, resulting in errors like PP's *mint, rug* → "rint, mug" and the similar errors shown by normal subjects in repeating strings of nonsense words. As discussed earlier, PP retained an excellent digit span throughout the period of follow-up, which we interpret as showing normal auditory-verbal short-term memory capacity. Any failure on the word-list repetition task cannot, therefore, be due to impaired working memory.

As mentioned earlier, the technique of comparing immediate recall for *known* and *unknown* words was introduced by Warrington (1975), in a study of two patients (EM and AB), and was also employed by McCarthy & Warrington (1987) with one patient (NHB). These three previously studied patients had very similar characteristics to those in our investigation (though none of the three was as severely impaired as PP), but produced rather different patterns of performance in this paradigm. In brief, none of these patients showed PP's significant advantage for known relative to unknown words in repeating lists of unrelated words; a number of possible explanations for this discrepancy are discussed in a companion paper (Patterson, Graham, & Hodges, 1994). By contrast, and obviously more in line with our results, McCarthy and Warrington's patient NHB showed a marked decrement in repetition of sentences containing an unknown word and produced many "phoneme migration" errors of the type shown in PP in word-list repetition.

### *Reading Aloud*

The hypothesis regarding the phonological lexicon was extended to the domain of reading in a paper by Patterson and Hodges (1992). By their argument, one consequence of the loss of semantic binding on lexical representations should be the reading disorder known as surface dyslexia. When invited to read words aloud, surface dyslexic patients perform well on regular words (i.e. words that embody a typical pattern of spelling-sound correspondences, such as *mint*) but very much less well on exception words (like *pint*) that deviate from regular patterns. Errors to exception words are usually straightforward "regularisations" such as *pint* pronounced to rhyme with "mint."

Accordingly, before we gave PP even a single word to read, we made the strong prediction that she would show a surface dyslexic pattern of reading. When presented with the word *have*, however, instead of producing the predicted regularisation error "haive," PP responded "I can't read"; she then proceeded to demonstrate the correctness of her insight (when we convinced her to try) by saying "N . . . A . . . Y . . . no, not Y . . . V, E; oh, nave." This is the pattern of reading associated with a different form of acquired alexia, pure alexia, in which patients (1) have a varying degree of impairment in recognising letters, and (2) tend to read in a letter-by-letter fashion. As it turns out, however, our prediction was not wrong. Classic pure alexic patients can, once they have correctly identified the component letters of a word, translate these into a correct pronunciation whether it is a regular or an exception word; but (as demonstrated by Patterson & Kay, 1982, and others) some pure alexic patients are also surface alexic; and this turned out to be true for PP. Her attempts to identify the letters in written words were so slow and errorful that we were forced to abandon standard forms of reading tests; but like all pure alexics, PP could "read" words (that is, translate them from a sequence of letters to a whole-word pronunciation) if she was presented with spoken letter names (e.g. "what does M, I, N, T spell?"). PP's performance in this identification from oral spelling, on a list of 252 words (126 pairs of regular and exception words matched for frequency and length) showed (1) a massive advantage for regular as compared with exception words, (2) a large frequency effect (she correctly pronounced 36% of the high-frequency exception words but only 8% of those low in frequency) and (3) a preponderance of regularisation errors on exception words (see Patterson & Hodges, 1992, for details). Asked "what does H, A, V, E spell?", PP did indeed reply "haive."

### *Comment*

The basis for the predicted association between semantic dementia and surface dyslexia is as follows: If phonological (and probably also orthographic) lexical representations have lost a significant degree of integrity because of the lost link to meaning, then the translation of orthography to phonology required for reading aloud will be based primarily on sub-word-sized elements. (Once again, for the sake of argument, let us suppose that onsets and rimes are the critical sub-word elements; this over-simplification is not troublesome because the inclusion of even smaller units—although making the explanation more cumbersome—actually makes the argument stronger rather than weaker.) Computation of phonology from orthography based on onsets and rimes will standardly yield the correct pronunciation of a regular word like *mint*, because the most typical pronunciation of *int* is the appropriate one for *mint*; by exactly the same logic, if the *int* portion of the written word *pint* is pronounced without reference to its onset, a regularisation error will result.

Virtually all patients with a relatively pure progressive disorder of semantic memory who have been tested on appropriate reading tasks have displayed this pattern of surface dyslexia, though as far as we know, PP is the first to show the combination of surface and letter-by-letter reading.

## Structural Descriptions of Objects

The ability to recognise possible objects as real objects is thought to depend upon the operation of a module containing the "stored structural descriptions" of familiar objects. This module is usually conceived of as separate from the semantic system per se. Hence it is possible that disintegration of semantic memory should have no impact on ability to perform an object decision test; indeed, Humphreys and colleagues (Riddoch & Humphreys, 1987; Sheridan & Humphreys, 1983) have argued for just this position. Alternatively, one might predict that structural descriptions of objects share with phonological representations of words the fact that, in normal processing, they are always active in conjunction with the relevant semantic representations. When one hunts for a comb on the cluttered bathroom shelf, activation of the comb's structural description and of the set of semantic features concerned with tidying one's hair are mutually active and reinforcing. It therefore seemed an open question whether a patient like PP, with profoundly disrupted semantic knowledge about most common objects (both living and man-made), should be able to discriminate possible and real objects from possible but unreal ones.

In August 1990 we administered the Object Decision Test of Riddoch and Humphreys (1987); half of the 64 pictures in this test are line drawings of real objects (e.g. a pair of scissors, a tiger) whereas the other half are chimeric objects (e.g. the body of a tiger with the head of a mouse, the blades of a pair of scissors on the handle of a screwdriver). The total set is subdivided into an easy set and a hard set. Control subjects perform well in discriminating between real and unreal objects even on the hard set; they perform almost perfectly on the easier set (mean control score for the easy drawings = 31.2/32 = 97.5 ± 3.7%). PP's performance on the easy set was 9/32 (28%). Her score was below chance due to the fact that she accepted all of the unreal objects as real, and rejected many of the real objects; although it is of note that all nine correct responses were appropriate acceptances of real objects.

### *Comment*

Since we know (from PP's performance on the object *matching* test) that her high-level visual processing skills were intact, her complete failure to discriminate between real and non-real objects is presumably to be attributed to impaired structural descriptions of familiar objects. We further hypothesise that these structural descriptions *cannot* function normally when there is profound loss of meaning. As noted earlier, in normal processing, object representations are always activated in conjunction with the meanings/functions of those objects; we suggest that when the meaning is lost, object representations lose their integrity.

## DISCUSSION

A number of patients with selective and progressive loss of semantic memory have been described previously; but the patient reported here, PP, is unique in the severity of her impairment. For example, the three patients studied by Warrington (1975), who were significantly impaired on all tests of semantic knowledge, each managed to name some items (either from picture or verbal description) and to give adequate if simple definitions of many high-frequency words; by contrast, PP never named or defined a single item correctly, even at the beginning of our study. Furthermore, despite PP's already profound deficit on initial assessment, follow-up over the next two years revealed further decline, culminating in virtually complete absence of any "declarative" semantic knowledge. Although this syndrome has a strong component of progressive aphasia, it is more fully characterised by the term semantic dementia, which conveys a pervasive semantic deterioration disrupting not only language but also conceptual knowledge, factual memory, and object and face recognition. The neurological and general neuropsychological details of five patients with semantic dementia, including PP, have been reported elsewhere (Hodges et al., 1992a). The purpose of the present paper, on the most severe of these five cases, was to report some detailed longitudinal data reflecting the dissolution of semantic memory, and more particularly to consider the impact of this dissolution on other cognitive processes. The discussion will focus firstly on one key aspect of the semantic memory impairment; secondly, upon components of the cognitive system that appear to function quite independently of semantic memory; and thirdly, on several components which, although regarded as independent modules, seem to be disrupted in a predictable fashion by loss of semantic memory.

Our tests of semantic knowledge did not, at any stage, reveal any category-specific preservation within PP's semantic memory nor any modality-specific preservation in her access to meaning; but it is of course possible that selective deficits of this type might have been present at an earlier phase of her disease. The deficit was profound, and impervious both to type of stimulus (words, sounds, pictures or real objects) and to mode of response (speech, category sorting, pointing, or gesture). On initial testing, however, when asked to sort pictures (from the semantic test battery of Hodges et al., 1992b), PP achieved perfect performance at the superordinate level (living vs. man-made things), impaired but above-chance success at a category level (e.g. land animals vs. birds), and complete absence of ability to classify at an attribute level (e.g. native vs. foreign animals). This pattern of results is similar to that observed by Warrington and co-workers (Warrington, 1975; Warrington & Shallice, 1984) in patients with selective loss of semantic knowledge. Follow-up for the first year revealed a rapid decline at the level of category membership but reliably flawless classification into living and man-made groups. Then, in the second year, even this most basic discrimination declined monotonically to chance. Unfortunately, we did not do longitudinal testing of PP's ability to sort the same corpus of items presented as words, but this would undoubtedly have been of considerable interest.

To perform the picture classification task, at any level, it is essential to understand both the general concept on which the classification is based (e.g. something about what distinguishes living from non-living things) and also the concept as applied to each item in the test. Thus, it cannot be argued that PP's perfect performance at the first level and partial ability at the second level of the test merely indicated a preservation of the overall concepts. That is, she must have been deriving information from the pictures that was sufficient to permit reliable classification about animacy and some degree of discrimination about category membership, but was insufficient to identify more specific attributes. This finding can be interpreted in at least two ways. First, it is possible that these levels of classification actually correspond to the hierarchical fashion in which semantic knowledge is organised (Collins & Quillian, 1969). Thus, for example, one's knowledge that a tiger is a land animal would be subordinate to knowing that it is an instance of the more general class of living things, and the knowledge that (in Britain) it is a foreign rather than a native animal would be even more peripheral in the hierarchy. According to this account, which in one form or another is the interpretation that has usually been applied to patients with semantic memory loss (e.g. Warrington, 1975), the pattern of deficit found in PP would imply that the disease process gradually prunes the hierarchical "knowledge tree," starting with the most specific twigs and only later reaching the more major branches.

Alternatively, one can think in terms of a more distributed framework, in which a semantic representation consists of a set of semantic features or nodes. By this account, higher levels of the putative tree are emergent properties reflecting many overlapping features of physically or functionally similar objects. A gradual loss of features from this network should blur a discrimination based on attributes before it disrupts the ability to make higher-level judgements, because the latter can be supported by a much reduced subset of features. To classify a tiger as foreign, large, or fierce (but a domestic cat as native, small, or tame) requires specific features; merely knowing something general about the body shape of both animals would probably be sufficient to discriminate them from birds or fish (for a further discussion of this issue see Rapp & Caramazza, 1993). We prefer this second account, which is more in keeping with current network models of semantic knowledge organisation (Allport, 1985; Farah & McClelland, 1991).

Another way of characterising the difference between these two accounts is as follows. According to the first, when presented with a picture of a tiger, PP might somehow still have recognised it *as a tiger*; but her only remaining knowledge about tigers would be at the highest (animate/inanimate) level. According to the second account, PP may have known nothing specific to tigers at all; her reliable assignment of this picture to the class of living things indicates not that she knew that *a tiger* was a living thing, but only that she could extract some non-tiger-specific information from the picture, information which shared more features with a general (and no doubt, in her case, impoverished) concept of living things than it shared with a concept of man-made objects. The fact that PP could not perform object decisions at a level above chance would seem to support this latter point: If there is a specific stage of object processing that corresponds to "recognition," this seems likely to be the stage at which a stimulus makes contact with the stored structural description for that object. Since PP could not discriminate between the appropriateness of a tiger's body with a tiger's head and a tiger's body with a mouse's head, it seems hard to argue that she could somehow still recognise tigers.

Turning to the aspects of cognition that appear to function normally in the presence of grossly impaired semantic memory, we have presented evidence that some basic syntactic processes, non-verbal problem solving, visuo-spatial skills, the perceptual processes necessary for judgement of object constancy, and short-term memory for auditory-verbal and spatial material are all cognitive modules that are relatively independent of semantic memory. The *content* of PP's spoken language obviously deteriorated dramatically over the course of her illness, but the syntactic form in which she expressed this diminishing content was always well structured, although it must be acknowledged that, by the end of the follow-up period, she was producing only a very limited repertoire of stereotyped "frozen" phrases. Early in the course of our investigations, we were also able to demonstrate reasonable levels of performance on two standard tests of syntactic comprehension, the Token Test and the TROG (Bishop, 1983), provided that allowances were made for her very restricted knowledge of referential meaning. Furthermore, in an on-line test developed by Marslen-Wilson and Tyler (1980), PP showed a normal advantage (in word-monitoring RT) for syntactically structured sentences as compared to random strings of words, plus a normal advantage for later target positions in structured sentences. This pattern of relatively well-preserved syntactic processing in the presence of severe impairment of semantics has also been observed in other cases of progressive semantic memory loss (Breedin & Saffran, Note 1; Schwartz et al., 1979).

One of our most striking results was the dissociation between PP's performance on an object-matching (or unusual views) task of object identity and on an object decision test of real vs. non-real objects. In Shallice's (1988) terms, this was a classical dissociation: That is, PP's performance was at chance level in object decision but completely normal in object matching, at least when first tested in August 1990. The investigation of object matching or constancy by the use of unusual views tests was initiated by Warrington and Taylor (1973; 1978), who demonstrated that patients with right posterior cerebral hemisphere damage had particular impairment on such tasks, due to a deficit in what they termed pre-semantic perceptual categorisation. Humphreys and Riddoch (1984; Humphreys, Riddoch, & Quinlan, 1988) extended Warrington's work and proposed two independent means of achieving object constancy; one that makes use of an object's distinctive features, and the other that utilises its principal axis of elongation. They showed selective impairment when the latter variable was manipulated—by presenting unusual foreshortened views of the objects—in a group of patients with right hemisphere damage, and the opposite pattern—selective impairment on a minimal-features version of the test—in an agnosic patient, HJA, with bilateral occipital damage. PP performed normally on both the foreshortened and minimal-feature versions of Humphreys and Riddoch's test, despite being unable to identify any of the objects represented. It should be noted, however, that the relationship between tasks of object constancy (e.g. unusual views) and object familiarity (e.g. the real–unreal object decision test), and their relative dependence on a putative stored structural system is controversial. By some accounts (see McCarthy & Warrington, 1990a), the stored structural descriptions of known objects are the basis of object constancy. In other models, the structural level of processing is separate and subsequent to the derivation of object constancy (Humphreys & Riddoch, 1984; Riddoch & Humphreys, 1987; Schwartz & Chawluk, 1990). Our results strongly suggest that the kind of object-centred analysis that is essential for recognising that two different views represent the same object is a separate process from knowing what the object is, or even whether it is familiar. Familiarity (as assessed by object decision) depends upon intact structural descriptions of known objects, whereas object constancy can be achieved without recourse to these representations. It is possible, although unlikely, that the single dissociation observed here may have arisen through differential task difficulty. Further confirmatory studies are required before definite conclusions can be reached.

It is clear that other high-level visuo-perceptual processes are also relatively independent of semantic knowledge. PP's performance on Benton's Judgement of Line Orientation and Facial Recognition Tests (Benton et al., 1983) was within the normal range for her age; and she was able to copy the Rey Complex Figure almost flawlessly. That her excellent copying relied on immediate, and non-conceptual, perceptual abilities was dramatically apparent from the following contrast: Asked to reproduce, from copy, either a complex meaningless pattern like the Rey Figure or a simple meaningful pattern like the outline drawing of a fish, PP performed both easily; but when we showed her the fish for a minute and then removed it before asking her to draw it, she could not do so at all, presumably because she had no higher-level conceptual representation on which to base a drawing of the object.

In common with other patients suffering from semantic dementia (Hodges et al., 1992a), PP's performance on a non-verbal reasoning test, Raven's Coloured Progressive Matrices, remained within normal limits for her age over the initial 18 months of follow-up. These data argue convincingly for a separation between semantic memory and reasoning ability based on the manipulation of non-verbal material. In addition, we have evidence for the preservation of both auditory-verbal and spatial short-term memory (Baddeley, 1986) in the form of PP's excellent digit and block-tapping spans. In contrast to the eventual decline on virtually all other tests observed after two years of sequential study, PP's forward and reverse digit span remained within the normal range. Working memory can clearly function in the absence of the semantic system. PP's performance does, however, suggest that auditory-verbal working memory is not impervious to the content on which it must operate. Although she had a normal (indeed superior) digit span, and a reasonable span for other words still within her vocabulary, she had difficulty in repeating common words whose meanings she no longer understood (discussed further later). We did not perform extensive tests to investigate her "comprehension" of numbers; but given her excellent span for numbers, it is noteworthy that she could still perform simple arithmetic operations at a time when her semantic memory for most objects and concepts was grossly impaired; and on latest testing, when she could produce no meaningful propositional speech at all, she could still count aloud.

The longitudinal data show that, although performance on non-semantic tests was initially intact, it did eventually decline; for instance, after approximately 18 months there was a dramatic drop-off in PP's performance on the Raven's Coloured Progressive Matrices and Rey Complex Figure Test. We assume that this extension of the cognitive deficit reflects a parallel spread of the pathological process beyond the brain regions concerned with semantic memory. The syndrome of progressive loss of semantic memory for which we have adopted the term semantic dementia (see Hodges et al., 1992a; Snowden et al., 1989), is consistently associated with selective and often asymmetric temporal-lobe abnormalities. This is apparent on structural, and particularly on functional (PET or SPECT), brain imaging. It is interesting to note that, in the PET results for patient PP, the posterior portion of the superior temporal gyrus (i.e. Wernicke's area) appeared to be spared, implying that this area is not primarily involved in the semantic aspects of language processing. These findings are in keeping with reports of selective impairment of semantic memory in the context of *non-progressive* brain damage, for instance following herpes simplex virus encephalitis or traumatic brain injury (e.g. Pietrini et al., 1988; Warrington & Shallice, 1984; Wilson, in press). Thus, there is converging evidence implicating left-temporal neocortical areas as the region most critically involved in the maintenance of semantic memory. The pathological basis of semantic dementia remains uncertain, but we have grounds for believing that it is unlikely to be Alzheimer's disease; many of the clinical, neuropsychological, and radiological findings point to a diagnosis of Pick's disease (for fuller discussion of this aspect, see Hodges et al., 1992a).

We turn now to a consideration of those processes which, though traditionally regarded as functionally distinct from semantic memory, were impaired in PP. We have investigated two cognitive domains: (1) the phonological representations of words, as assessed by tasks of repetition and reading aloud, and (2) the stored structural descriptions of objects, as assessed by tasks of object decision. Our theoretical interpretation of the first of these is discussed in detail elsewhere (Patterson & Hodges, 1992; Graham, Hodges, & Patterson, 1994; Patterson et al., 1994), so we shall confine ourselves here to a brief overview.

The integrity of the phonological representations used in speech production was investigated using a strategy devised by Warrington (1975), in which the subject is asked to repeat strings of "known" and "unknown" real words. PP showed both a dramatic advantage for immediate repetition of known relative to unknown strings and a preponderance of phoneme-migration errors in this task. We interpret these results by proposing that representations in the phonological lexicon, which in normal processing are in constant interaction with representations of meaning, actually depend in part for their integrity on this link to meaning. The elements of phonological representations themselves are not corrupted by loss of meaning, which is why PP (unlike, for example, a severe Wernicke's aphasic) could repeat single words or nonwords without difficulty. It is producing the elements in the right combinations (if there is more than one set) that suffers when meaning no longer specifies "mint" as an appropriate combination of elements but "rint" as an inappropriate one (cf. PP's repetition error *mint, rug* → "rint, mug"). PP's surface dyslexia can also be understood by this notion of reduced coherence in phonological (and orthographic) representations due to loss of meaning. We argue that fragmented lexical representations will force the translation from orthography to phonology to be based primarily on sub-word sized elements. As the most common translation of the segment *int* is /ɪnt/, *mint* will be read aloud correctly but *pint* will tend to be regularised. The fact that PP was a pure alexic (and poor letter-by-letter reader) does not alter this interpretation of her surface reading pattern, though (1) it forced us to use identification from oral spelling rather than standard printed words to investigate her "reading," and (2) the pure alexia is of interest in its own right.

Our hypothesis about the impact of semantic loss on structural subsystems that are in constant communication with semantic memory also applies to the domain of object recognition. We employed the object decision test developed by Riddoch and Humphreys (1987) to investigate PP's structural description system. The latter system can be defined as the representations of perceptual knowledge about familiar objects that mediate between peripheral visual analysis and semantic memory. PP was, even on initial testing, essentially at the level of chance in the object decision test. Since she was able to classify similar drawings of real items into living and man-made categories, one cannot argue that her failure on the object decision test was due to a general inability to classify pictures. If (as argued by most theorists—see for example McCarthy & Warrington, 1990a; Sheridan & Humphreys, 1993) the structural description system is a separate processing module rather than an integral component of the semantic system, then results from PP (and from other patients with semantic dementia who have also demonstrated marked impairments in object decision: Hodges et al., 1992a) suggest that the two are separable but not functionally independent. Our tentative interpretation of this functional interdependence is precisely parallel to our account of the relationship between semantic memory and phonological representations. The phonological string "mint" is no better than or different from the string "rint" except in that the former is familiar and meaningful whereas the latter is not. Likewise, the combination of a deer's body with a deer's head is no better, in principle, than a deer's body with a dog's head; again, the difference lies in the fact that only one of these combinations of familiar elements has a corresponding representation in semantic memory. After 68 years (most of these normal and healthy) of seeing real dogs and real deer but (presumably) never a combination of the two, PP apparently found the false combination as plausible as the real object. This seems to us a dramatic demonstration that structural descriptions do not remain intact when they lose communication from semantic memory.

Thus far, our investigations of the impact of semantic memory loss on structural "satellite" systems have been restricted to these two domains of phonological representations for speech and visual representations of objects; but we hypothesise that cognitive modules serving parallel roles in other domains will be similarly affected. PP's performance on some informal tests of face recognition provides hints of this in the domain of face processing. If her profound deficit in semantic knowledge about people had no further consequences, then, according to contemporary models of face processing (see Bruce & Young, 1986; Ellis & Young, 1991; Hodges, Salmon, & Butters, in press), she should still have been able to discriminate between famous and non-famous faces. However, she failed to recognise any famous faces, not only their identities but also their familiarity, despite the fact that her performance on a face matching test was within the normal range. The input and output systems surrounding semantic memory clearly warrant further exploration in this extraordinary syndrome of semantic dementia.

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