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Relearning and retention of verbal labels in a case of semantic dementia

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Background: Previous studies looking at relearning and retention of word labels in people with semantic dementia have shown some improvement in naming immediately after the period of learning but this has not usually been maintained. Studies have also shown rigid learning of names, in the order of presentation and to the picture exemplars only, with no generalisation of learning.

Aims: This study aimed to explore relearning of a small vocabulary set in a person with semantic dementia (CUB) and to examine her ability to generalise this learning. In addition, it aimed to find out how long the learning persisted after therapy was completed given that semantic dementia is a progressive disorder.

Methods & Procedures: A single-case design was used where CUB was asked to learn 28 words while a further 28 were left as controls. A “look and say” method was used daily for 1 month. As well as examining learning of the therapy and control set, CUB was asked to name 168 other exemplars of the learning set to see whether there had been any transfer of her learning from the therapy set.

Outcomes & Results: CUB not only relearned a set of picture names but retained these without deliberate practice over a 6-month period. She was also able to generalise this learning to other visually similar exemplars in testing and in daily use. The maintenance of relearning was achieved despite severe deterioration in her semantic memory.

Conclusions: Possible reasons are explored as to why CUB was able to relearn and retain these words and why this may differ from all previously reported cases. Differences in amount of time spent relearning, number of items learned, therapy methods, the severity of semantic memory impairment, the degree of atrophy, and the behavioural profiles of people with semantic dementia do not provide adequate explanations for our individual’s differential ability to retain her learning over 6 months. The most plausible explanation is that the person with semantic dementia generalised her learning to her everyday speech and this provided the source of maintenance for the relearned names.

Keywords: Semantic dementia; Anomia; Relearning; Generalisation; Maintenance.

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Semantic dementia is the temporal lobe variant of fronto-temporal dementia in which there is progressive but circumscribed bilateral atrophy of the anterior, inferolateral temporal lobes. This damage produces a gradual and eventually profound deterioration in semantic memory (Hodges, Patterson, Oxbury, & Funnel, 1992). The impairment affects both receptive and expressive skills and verbal as well as nonverbal modalities (Bozeat, Lambon Ralph, Patterson, Garrard, & Hodges, 2000b; Lambon Ralph & Howard, 2000). The syndrome is relatively homogeneous, with variations dependent on the relative atrophy of the left and right temporal lobes (Lambon Ralph, McClelland, Patterson, Galton, & Hodges, 2001; Snowden, Thompson, & Neary, 2004). A number of characteristics have been shown to be constant within semantic dementia (Rogers et al., 2004). For example, frequency and familiarity determine which concepts are better retained. General attributes (e.g., the legs of a zebra) are retained better, while item-specific information is more vulnerable (e.g., the stripes of the zebra), so that distinguishing between similar concepts becomes harder.

The amodal semantic impairment found in semantic dementia results in a profound anomia (Lambon Ralph et al., 2001). Indeed this is the most common presenting symptom in this patient group and causes considerable anxiety. Even people with extremely mild comprehension impairments tend to have significant word-finding difficulties, which accelerate over the early course of the disease (Lambon Ralph et al., 2001). Both clinically and academically, therefore, it is imperative to explore methods and techniques designed to restore, or at least maintain, a core activities of daily living (ADL) vocabulary in this patient group.

Despite the extensive literature dedicated to establishing the nature of the deficits in semantic dementia, studies looking specifically at interventions for these word-finding difficulties number only six to date (Frattali, 2004; Graham, Patterson, Pratt, & Hodges, 1999, 2001; Jokel, Rochon, & Leonard, 2002, 2006; Snowden & Neary, 2002). A summary of cases in each of these papers is provided in Table 1, along with scan details and background information. In each study, people with semantic dementia have benefited in the short-term from mass practice of selected concrete concept names. However, the retention of this learning over the longer term (measured between 2 weeks and 6 months after learning) without continued practice has been minimal or non-existent. Furthermore, the ability to retain words has been shown to rely on the degree of semantic degradation present at the start of the therapy programme (Snowden & Neary, 2002). If some meaning remains, then retention of the re-learned word is longer but this partial conceptual knowledge does not prevent its eventual loss (Jokel et al., 2002, 2006). Where little meaning remains, it is very hard for the person with semantic dementia to relearn the corresponding label, let alone retain it without practice. Some specific details about the five existing case studies are reviewed below.

Graham et al. (1999) described the case of DM, a 59-year-old surgeon who had developed a method of note-keeping to retain words that he knew were disappearing from his vocabulary. He was meticulous in this method and would practise the lists he had made for himself over several hours a day. At the time of the study, DM's semantic abilities and naming skills were still relatively unimpaired. He scored 49/52 on the three-picture version of the Pyramids and Palm Trees Test (PPT, Howard & Patterson, 1992) and 71% on the Cambridge picture-naming test (Hodges et al., 1992). DM was given 100 words from four different categories to relearn while

TABLE 1
Summary of cases with semantic dementia where relearning has been undertaken, method used, and short- and long-term effects

<i>Case</i>	<i>Reference</i>	<i>Age</i>	<i>Scan</i>	<i>Background</i>	<i>Semantic impairment</i> ¹	<i>Method</i>	<i>ST effects</i>	<i>LT effects</i>
DM	Graham et al., 1999 (& 2001)	59	L temporal lobe atrophy, involving the pole and to smaller extent inferior region of mid and posterior temporal pole.	Surgeon	Mild 49/52 (PPT)	List learning within categories.	√	x
KB	Snowden & Neary, 2002	64	Severe atrophy of temporal lobes R > L. Lesser involvement of medial temporal lobes.	NR	Moderate to severe 25/52 (PPT)	Picture/spoken and written word labels.	x	x
CR	Snowden & Neary, 2002	57	Selective atrophy L temporal lobe (MRI) L > R.	NR	Moderate to severe 29/52 (PPT)	Picture/spoken and written word labels with autobiographical linking.	√	partial
AK	Jokel et al., 2002 & 2006	63	Bitemporal atrophy, L > R. Some atrophy of L ventral frontal regions.	Arts officer	Moderate 37/52 (PPT)	Picture/spoken and written word labels.	partial	x
	Frattali, 2004	66	Focal atrophy inferior medial aspect of L temp lobe relative sparing of hippocampus.	Military	Moderate to severe	Pictures in conversation.	partial	x

¹As measured using Palm Trees and Pyramids (Howard & Patterson, 1992) or equivalent. ST = short term (immediately post-therapy and up to 1 month post-therapy). LT = long term (6 months). NR = not recorded. Partial: indicates that the participant was able to learn the verbal labels for some of the items.

another four categories were kept as controls. He was asked to read and say the target words aloud for 30 minutes each day, for a total of 2 weeks. He was then re-assessed by generating as many items as he could for each of the four therapy categories and for the four control categories. Although DM showed immediate benefits from this learning method, in that he was able to generate significantly more category items than at baseline, he was not able to generate any new exemplars for that category. He also provided the list in almost exactly the same order as he had learnt it during therapy. DM showed a surprising improvement in one of the control categories (makes of car) and when this was investigated further, it transpired that he had been looking at these in a book over the 2 weeks and so, in effect, had also been relearning these items without the examiners' knowledge. After 8 weeks without practice, DM's performance returned to baseline levels. A follow-up study (Graham et al., 2001) of DM's retention of the learned categories was carried out 2 years later. DM had practised the lists during the 2 years and was able to recall these, despite deterioration on tests of semantic knowledge. These studies highlighted four characteristics that have been repeated across other investigations (see below): (1) persistent and regular practice seems to allow relearning to occur in semantic dementia; (2) maintenance of the relearning requires constant practice; (3) performance rigidly reflects item order; (4) there is little evidence of generalisation from target items to other concepts or exemplars.

Snowden and Neary (2002) studied two cases whose semantic and anomic difficulties were more severe than DM. KB was a 64-year-old woman who scored 25/52 on the three-picture version of the PPT (i.e., her performance was at chance) and was unable to name any pictures in the Boston Naming Test (BNT; Kaplan, Goodglass, & Weintraub, 1983). Information on KB's semantic knowledge of 60 line drawings from the Snodgrass and Vanderwart corpus was collected by asking her to name the picture and provide definitions both to the picture and the spoken word, and spoken word-to-picture matching. From these 60 items, 20 pictures that she failed to name were selected. KB was unable to demonstrate knowledge of 10 of the selected items and showed partial knowledge for the remainder. The learning procedure consisted of a baseline assessment of the 20 items during which KB attempted to name the pictures. KB was then shown each picture and its written label and was asked to read the word aloud while concentrating on the picture. The examiner then repeated the word aloud. All 20 pictures were then retested. This procedure was carried out three times in total. KB was then given a 2-week break during which she did not look at the materials and then two further learning sessions. She was retested 4 months later. KB learned a few words after the 2-week break, though not sufficient to show increase in naming accuracy over baseline. It is noteworthy, however, that items that were learnt came from the set for which she had shown partial semantic knowledge at the start.

Snowden and Neary (2002) contrasted this case with CR, a 57-year-old woman whose semantic and anomic difficulties were also severe. She scored 29/52 on the three-picture version of the PPT (again at chance) and 4/60 on the BNT. A similar procedure to that used for KB was used to establish a learning set of 20 items, although this time CR showed no knowledge on measures of naming, definition, and word-to-picture match for any of the 20 items. The learning trials were similar to those of KB in that she was shown the picture and written label, and asked to read the word aloud. The learning procedure was augmented in two ways: she was asked to provide descriptive information about the meaning of the word and to make links

between the picture and objects in her own home/environment. CR then looked again at the picture and written label, this time with a spoken commentary from the experimenter about the item, using the definition and home environment information provided earlier. For example, for the item “duck”, she was reminded that she had an ornamental duck on her wall and that there were ducks in the pond. Following this learning session, CR was provided with a self-study booklet for the 20 items that she had to look at, read, and say. CR practised for 20 minutes each day for 3 weeks and was faithful to the practice regime. When she was unable to remember a name she asked her husband for the semantic link, which he would then provide. CR’s naming ability was perfect (20/20) both immediately and 60 days later. Her score dropped to 13/20 when tested 6 months later. At the 60-day assessment and at 6 months, the 20 pictures were also tested using random presentation order and with a different colour background. These changes produced a drop in CR’s performance (15/20 and 8/20 respectively). These results imply a strongly context-dependent learning mechanism underlies these gains.

Jokel et al. (2002, 2006) described the case of AK, a 63-year-old woman who was an arts officer and a keen musician. Her semantic abilities and naming skills were moderately severe (she scored 37/52 on the PPT and 5/60 on the BNT). AK was asked to name, on two occasions, the 230-picture set from the Peabody Collection (Dunn & Dunn, 1997). Her comprehension of the items was tested using word-to-picture matching of the items. A set of 180 items was established, subdivided into three sets: 60 that she named and comprehended (+N+C), 60 that she did not name but did comprehend (–N+C), and 60 that she neither named or comprehended (–N–C). These sets were then further divided into treatment and control sets. The 30 pictures of each subset were placed on card with the written label and a brief description of the item (previously provided by AK) on the reverse. AK looked at the picture and read aloud the label and description of the item. She carried this out at home, practising for half an hour per day for 6 days. On the seventh day she was tested on the 30 items intermingled with 30 control items. Each of the three subgroups was treated for 1 week (a total of 3 weeks) in a specific order: first +N+C; then –N+C and finally –N–C. Retest of all therapy and control items was carried out 1 month after the end of the last treated set and 6 months later. Three important results arose from this study. First, items that were practised were retained better at all stages than items which were not. Second, best results were achieved with those items where AK retained some semantic knowledge. Lastly, even with practice, some items were lost over time in the +N+C set.

Finally, Frattali (2004) described a 66-year-old retired military man who was asked to relearn 20 nouns and 20 verbs using an errorless (but effortful) paradigm in which the therapist and the gentleman engaged in conversation about pictures in which target items were displayed. Therapy lasted for 2 hours a week over a period of 12 weeks. Some improvement, particularly in noun naming, was evident immediately after the therapy but this was not maintained at the 3-month follow-up assessment.

In summary, the reported cases to date have generally shown some improvement in naming immediately after the period of learning but this has not usually been maintained. Where it has been maintained (Snowden & Neary, 2002) the apparent gains diminish if the testing order and/or the appearance of the test items is altered, suggesting that some gains reflect rote learning. Given the paucity of previous studies, this study chose to replicate what had been carried out in previous studies so

that comparison with other findings would be possible, and to extend previous findings by further post-therapy testing. In this study, various parameters that might influence relearning were explored with a person with semantic dementia, CUB. In striking contrast to the previously reported cases, CUB not only learned a set of picture names but retained these without deliberate practice over a 6-month period. She was also able to generalise this relearning to other visually similar exemplars in testing and in daily use.

CASE HISTORY

This study was approved by the local ethics committee of the University of Málaga. CUB was a 53-year-old married Spanish woman who had previously worked as a civil servant in the Justice Department until the onset of her illness. In 2002 she was signed off from work with depression. It was around this time that she began to have word-finding difficulties. She was first seen by our research group at the end of 2004, when her language was fluent with a tendency to talk garrulously such that, at times, it was difficult to interrupt her. Her speech was empty, with marked anomia, perseverations and the use of filler words such as “anyway”, “simply”, “precisely”, “do you understand me?”, “the topic is”, “I know about this”. She occasionally swore—something she had never previously been in the habit of doing. She complained of intrusive words that would come into her mind but for which she had forgotten their meaning (for example “níspero” [a soft fruit somewhat like a peach], “melocotón” [peach]). When this happened she would write them down. Her husband reported that she would then use them for a few days and then they would disappear again from her speech. Her husband also reported that she had some difficulty recognising old friends (although this was not the case with her immediate family). CUB had some insight into her problems, in particular, her progressive anomia. Comprehension deficits were apparent on examination; for example, when asked if she had a good appetite, she replied; “Appetite? I have forgotten. What is appetite?” An MRI scan from 2004 showed focal, bilateral temporal atrophy with a marked knife-edged shape in the left middle and inferior temporal gyri, but only mild atrophic changes in the right temporal lobe (Figure 1).

NEUROPSYCHOLOGICAL TESTING

Behavioural examination

CUB underwent a battery of neurological and neuropsychological tests at the end of 2004 and start of 2005. Her Mini-Mental Test score (Folstein, Folstein, & McHugh, 1975; Peña-Casanova, Gramunt, & Gich Fullà, 2004) was 24/30, failing the word recall and naming elements. General behaviour was examined via the Frontal Behavioral Inventory (Kertesz, Davidson, & Fox, 1997). She scored 21 (range: 0–36) on the negative behaviour scale and 15 (range: 0–27) on the disinhibition score, displaying apathy, inflexibility, loss of insight, semantic dementia, perseverations, irritability, inappropriateness, hyper-orality, and hyper-sexuality. The Leyton Obsessional Inventory (LOI: Cooper, 1970) was completed by her husband rather than CUB and so the interference and resistance scales could not be completed. The scores were very high (LOI symptoms: 45/46; LOI traits: 20/23) showing a number of

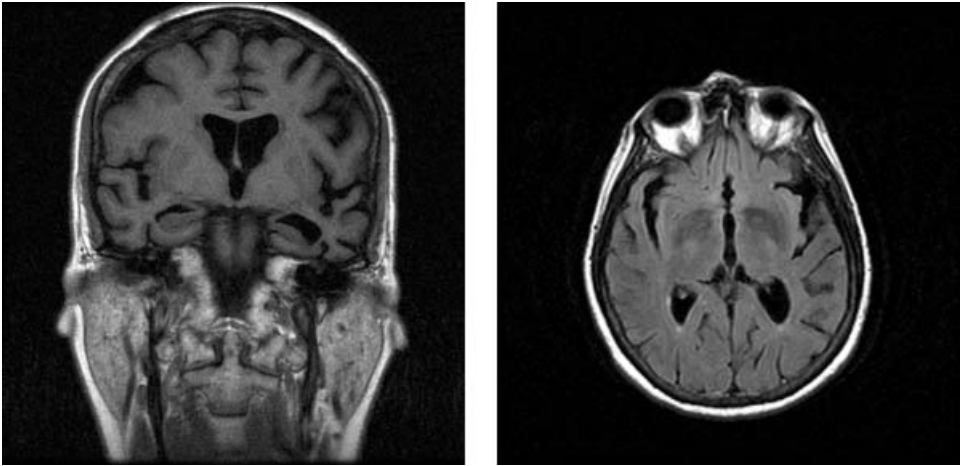


Figure 1. CUB: MRI coronal and axial slices. Note that the left hemisphere is shown on the right of the MR images.

obsessions (e.g., fear that her family would come to harm, distaste of any physical contact with people she did not know) and compulsions (e.g., the need to continuously check the gas, wash her hands). She also showed obsessional traits (e.g., the need to stick to a rigid timetable, sorting, ironing, and re-sorting her clothes continuously). The history provided by her husband suggested that some of the behaviours might well have been related to premorbid obsessive-compulsive personality traits. Other features are consistent with those found in semantic dementia and frontotemporal dementia more generally (e.g., clock-watching etc., Bozeat, Gregory, Lambon Ralph, & Hodges, 2000a; Snowden et al., 2001). CUB also reported that words would come into her mind unheeded. Some of these words were specific to her previous lifestyle (for example, administration, finance, and judicial vocabulary) while the rest were names of body parts and bodily functions as well as food and household items. There were also some animal names.

Cognitive testing

On the Raven's Coloured Progressive Matrices (Raven, 1985) CUB scored a total of 28/36, which placed her in the Grade II intelligence category (75th %ile). Her copy of the Rey figure (Osterreith, 1995) was perfect (36/36), an exact copy, while her immediate recall was severely impoverished (10/36, 3rd %ile), suggesting good visuospatial skills but severely impaired visual memory. On the Wechsler Memory Scale (Wechsler, 1976) she showed severe problems with logical memory (1/23), paired associate learning (3.5/21), and reproducing diagrams from memory (7/14). She showed severe difficulties with both semantic and letter fluency. She gave three animals in a category fluency task, plus one intrusion (a fruit "melón"). On the FAS (Borkowsky, Benton, & Spreen, 1967) she provided 11 correct responses. In addition, she gave three responses that were linked, not to the letter as required, but semantically ("sábana" [sheet], "cama" [bed], "manta" [blanket]). Her forward digit span (measured using EPLA/PALPA, Kay, Lesser, & Coltheart, 1992; Valle &

Cuetos, 1995) was 4. CUB retained her ability to read aloud and to spell accurately, as expected for languages with transparent orthography (Patterson, Okada, Suzuki, Ijuin, & Tatsum, 1998).

Face processing

Benton's Facial Recognition Test (Benton, Hamsher, Varney, & Spreen, 1978) assesses ability to match unfamiliar faces across different views. CUB scored 22 on the shortened form, which is equivalent to 46 on the full test version. Control performance is between 41–54 indicating that CUB had no difficulty with this task. In contrast, her ability to recognise and name famous faces was impaired. On this test, recognition is credited when the participant names the famous person or provides identifying information (for example, Juan Carlos Ferrero – ‘He’s a tennis player’). CUB’s recognition (12/56) and naming scores (2/56) were very poor in comparison to age-matched controls (naming: mean = 43.9/56, range 33–53).

Background language testing

On the Western Aphasia Battery (Kertesz, 1982), she gained an Aphasia Quotient of 66.2, which placed her in the category of anomic aphasia with particular difficulty in the naming subtest (1/20). In both spontaneous speech and picture description task, her speech was fluent but with a paucity of content words.

Phonological skills. Auditory discrimination was assessed using minimal word and nonword pairs from EPLA/PALPA (Kay et al., 1992; Valle & Cuetos, 1995). CUB showed a tendency to accept pairs as sounding the same (word test 28/28 same trials; 22/28 different trials, controls 27.68, $SD = 0.76$, nonword test 28/28 same trials, 23/28 different trials, controls 27.09, $SD = 1.24$). On both these tests of auditory discrimination, therefore, CUB showed a mild to moderate impairment. Although CUB’s hearing was not formally tested, this performance is unlikely to reflect a deficit in hearing acuity but is in line with other studies showing phonological difficulties in this client group, which are attributed to their semantic impairment (Jefferies, Jones, Bateman, & Lambon Ralph, 2005; Patterson, Graham, & Hodges, 1994; Patterson et al., 2006). On an assessment of auditory lexical decision from EPLA/PALPA, CUB performed at chance 48/160. CUB’s single word repetition was at ceiling for verbs (50/50), adjectives (50/50), and functors (40/40), and close to ceiling for nouns (119/120) and nonwords (77/80). This pattern of excellent repetition yet impoverished word recognition is typical of semantic dementia (Jefferies et al., 2005; Patterson et al., 1994, 2006). CUB was able to read aloud all the words and nonwords that she had been asked to repeat. This retained reading ability reflects the nature of Spanish orthography, which is transparent in the translation of orthography to phonology (i.e., there are no irregular words in reading, at least at the level of phonology, although some inconsistency is present for stress assignment).

Semantic memory. On the Pyramids and Palm Tree Test (Howard & Patterson, 1992) CUB scored 33/52 on the three-picture and 31/52 on the three written word version of the test, showing a severe difficulty in identifying semantic associations, irrespective of modality of testing (English-speaking controls made up to a maximum of three errors on this test, Spanish controls made a maximum of two

errors). On the spoken-word-to-picture matching task from EPLA/PALPA (Kay et al., 1992; Valle & Cuetos, 1995) she scored 15/40, making 11 close semantic, 8 distant semantic, 3 visual, and 3 unrelated errors (Spanish control mean 39.45, $SD = 1.67$).

CUB's semantic memory was reassessed 6 months after the end of the therapy programme (December 2005) when she scored 18/52 on the three-pictures version of PPT (Howard & Patterson, 1992), a performance that is at chance and significantly worse than at the start of the study when she scored 33/52 (McNemar, exact $p = .003$). By this time, her conversational speech had also deteriorated to stereotypical phrases and repetitive topics (such as her illness, her desire for a medicine that would help, etc.). She had begun to have behavioural difficulties such as wandering from home and kleptomania.

Naming skills. On the EPLA/PALPA naming by frequency test, CUB scored 13/60, 10 of which were high-frequency and 3 low-frequency items (which were, however, familiar and important everyday items for CUB).

Therapy method

From the 64 pictures that CUB failed to name in the Western Aphasia Battery and the EPLA/PALPA Naming by Frequency test, 56 were selected and divided into two sets of 28, one set for therapy and one set as a control. A list of these items and their translations are available in the Appendix. Both sets were matched on frequency, syllable, and phoneme length using BuscaPalabras (Davis & Perea, 2005). The mean values for both sets are set out in Table 2.

CUB's naming of the 56 items was re-measured prior to the start of therapy (baseline 2) when she was able to name 6 of the items in the control set and 2 in the therapy set (see Table 3).

CUB's errors at baseline were divided between no-response errors (60%) which she indicated in a number of ways ("I don't know this", "I have forgotten this", "I

TABLE 2
Mean frequency and length values for the therapy and control items

		<i>Frequency</i>	<i>Syllable length</i>	<i>Phoneme length</i>
Therapy items	Mean	36.26	2.61	5.86
	SD	57.92	0.69	1.69
Control items	Mean	36.31	2.50	5.50
	SD	52.14	0.69	1.35

TABLE 3
Naming accuracy for two baselines: Immediate post-therapy and at 1 and 6 months post-therapy

	<i>Baseline 1</i>	<i>Baseline 2</i>	<i>Immediately post-therapy</i>	<i>1 month post-therapy</i>	<i>6 months post-therapy</i>
Therapy items	0/28	2/28	28/28	27/28	23/28
Control items	0/28	6/28	5/28	6/28	0/28

used to know this”); descriptive information (25%), for example to the target “pram” she replied; “where you put children”; and superordinate errors (15%), for example to the target “pear” she replied; “it’s a fruit”. These errors are consistent with those shown by other people with semantic dementia (Jefferies & Lambon Ralph, 2006; Rogers et al., 2004).

A PowerPoint presentation was used for the 28 items, with each item pictured first, followed by the picture paired with its written label. In therapy, CUB tried to name the picture first but if she was unable to name it, she was then asked to move on to the page where the picture and written label were displayed together and to read aloud the name. The items were placed on a CD-ROM, which CUB looked at every day for 1 month, practising the items in the same order each day. During this self-practice, her husband reported that occasionally CUB would come and ask him what a word meant and he would show her the item or explain its meaning.

RESULTS

CUB’s naming of the 56 items was tested immediately after the month-long, self-directed home therapy and again at 1 month and 6 months post therapy. Following the month’s self-directed learning, CUB returned the CD-ROM to the researcher and did not continue to practise these words. During those 6 months CUB continued to take part in relearning other words selected by her husband and son.

In order to test her naming skills without the potential support of the original therapy order (see the introduction), the items were presented in a different order from that used in therapy and with the control items interleaved. Her last response was used to evaluate her answer. This was chosen as she would sometimes make a comment before naming—such as “this was on the computer”, “you didn’t put that on the computer”—and there were two items for which she produced “conduite d’approche” behaviour before naming the item (for example, to the picture of “hacha” [axe], she said “bacha, hacha” and “hucha, hacha” at 1 month and 6 months respectively). For two items “bañera” [bath] and “mesa” [table], she used a set phrase (which her husband had provided for her early in therapy when she had asked him for help). In order to access “bañera” [bath], she said “baño, bañera” [bathroom, bath] while for “mesa” [table], she said “mesa y silla” [table and chair]. The results of these assessments are shown in Table 3.

These results showed excellent relearning of the items immediately post therapy when she was able to name all the items. A comparison of scores at each of the three retest times (immediately post therapy, at 1 month follow-up, and at 6 months follow-up) showed a significant difference between the learned and control sets (immediately post therapy $\chi^2 = 35.07$, $df = 1$, $p < .001$, 1 month post therapy $\chi^2 = 28.99$, $df = 1$, $p < .001$, and at 6 months follow-up $\chi^2 = 35.07$, $df = 1$, $p < .001$). Naming improved significantly on the therapy set from baseline to immediate post therapy (McNemar, one-tailed, $p < .001$) and this significant difference was maintained at 1-month and at 6-month follow-up. A similar comparison of control items showed no significant difference (comparison of baseline with immediate post-therapy performance, McNemar, one-tailed, $p > .05$).

After 1 month without therapy, CUB named all but one item (“botón” [button]) although she recognised it as having been part of her therapy set. At 6 months follow-up, there were five items that she could no longer name (Table 4). Four of her five errors were the names of other items within the therapy set and related to the

TABLE 4
Targets and errors within the therapy set at 6 months follow-up, with error type

<i>Spanish target</i>	<i>English translation</i>	<i>Spanish error</i>	<i>English translation</i>	<i>Error type</i>
cama	bed	cubo	bucket	Within therapy set
globo	balloon	vaya	go	Phonologically related to another word in therapy set (vela – candle)
piano	piano	peine	comb	Within therapy set and phonologically related
sombrero	hat	botón	button	Within therapy set and semantically related
vaca	cow	dromodario	dromedary	Within therapy set and semantically related

target either semantically (“sombrero” [hat], – “botón” [button],) or phonologically (“peine” [comb] for “piano”). Although one error (“vaya” for “globo” [balloon],) was not in the therapy set, it was phonologically related to another therapy item (“vela” [candle]).

One important feature of conceptual knowledge is that it allows information about one stimulus to be appropriately generalised to other examples (McClelland & Rogers, 2003; Rogers et al., 2004). There is already some evidence to suggest that breakdown in conceptual knowledge produces under-generalisation in semantic dementia. Snowden, Griffiths, and Neary (1995) found that remaining conceptual knowledge (e.g., dog licence) did not generalise appropriately (e.g., to licences in general). Likewise in object use, Bozeat, Lambon Ralph, Patterson, and Hodges (2002) found that retained use for individuals’ own everyday objects only generalised to another example if the two were visually similar. For visually dissimilar objects, people with semantic dementia often failed to recognise that they were the same type of object. In an attempt to evaluate this possibility in the context of name relearning, CUB was given a further naming test in which six different examples of each of the 28 therapy items were presented. For example she was shown a single banana, a bunch of bananas hanging from a tree, diced bananas, etc. (Figure 2).

This assessment was carried out immediately after the therapy when CUB scored 155/168 (92%). Her excellent naming performance on these alternative exemplars indicates that her relearning did generalise. Inevitably, however, many of the

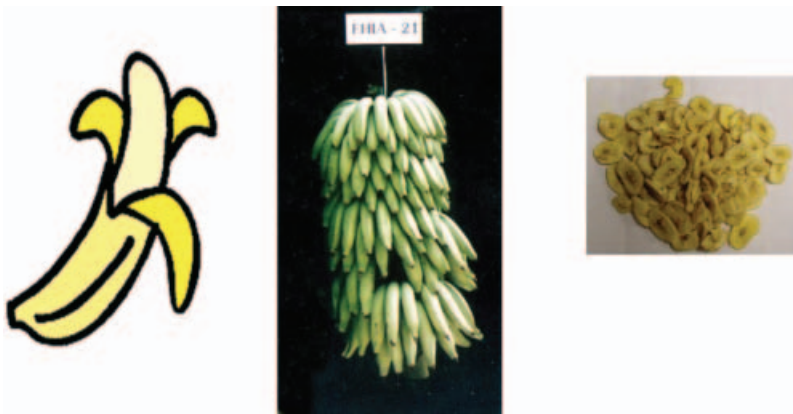


Figure 2. An example (“banana”) of the different exemplars of a therapy item used in 168-item test.

pictured alternatives were visually similar to the original pictures and thus (as Bozeat et al., 2002 found) the generalisation may have been based on visual similarity. In order to test this possibility, the 168 items were rated for typicality by 17 controls to obtain a mean typicality measure for each item. The typicality of the named and unnamed items was compared and CUB showed a significant effect of typicality, $t(166) = 3.349$, $p = .001$. For example, CUB made errors on the more atypically represented items, such as the cut bananas from Figure 2.

GENERAL DISCUSSION

Previous reports of relearning in cases of semantic dementia have shown poor retention of learning in both the short and longer term. Where learning has taken place, performance deteriorates when test items are presented in a different order or with different backgrounds (Snowden & Neary, 2002). We presented a case of semantic dementia, CUB, who had moderately severe anomia and semantic difficulties at the time of testing, which declined significantly during the therapy study. CUB was able to relearn and retain, over time, 28 names for common objects. Unlike previously reported people, she was still able to name these when they were presented in a different order from that used in therapy and when presented among control items she had not learned. Despite her excellent learning, there was some evidence of “rote” learning. Two items (“bañera” [bath] and “mesa” [table]) were learned as set phrases “baño, bañera” [bathroom, bath] and “mesa y silla” [table and chair] respectively. When CUB made an error she used the name of another item from within the therapy set, which may provide some additional indications of rigidity within the relearning—as has been found in previous relearning studies. When she was asked to name six other exemplars of each item she scored 155/168 (92%), making errors on those items that were not visually similar to the learned target. One of the most remarkable aspects of CUB’s abilities in relearning was the retention 6 months after relearning when, in all other respects, she had deteriorated severely. At that time, CUB’s performance on semantic tasks was at chance, her speech was empty and repetitive, she was unable to name items in the control set that she had previously been able to name, and she had begun to show behavioural problems such as kleptomania and wandering. Yet, without continued practice, she named 23/28 (88%) of the therapy set. This aspect of her performance contrasts with the previous studies. There are a number of possible explanations for such a striking difference and these are considered below.

Amount of therapy

The amount of time CUB was asked to spend on relearning was longer than in the three other cases. CUB was asked to look at the CD-ROM every day for a month. The other cases varied from 2 weeks (Graham et al., 1999) to 3 weeks (Jokel et al., 2002, 2006; Snowden & Neary, 2002). CUB’s husband reported that she did make an effort to look at the CD-ROM each day and, by day 3 of the month, she was able to recall the words without recourse to the written help on the next page. This suggests that she may have performed equally as well if she had been tested at 2 or 3 weeks after the start of her self-learning. It might equally be the case that the continued rehearsal of the items over the month, with the picture and written feedback, was

critical in the successful maintenance of her learning. This study cannot distinguish between these two possibilities.

Number of items

The number of items that CUB was asked to learn (28) differed from other studies, which varied from a maximum 100 words from within five different categories (Graham et al., 1999) to 20 autobiographical items (Snowden & Neary, 2002). The closest in set size to CUB's 28 items was AK (Jokel et al., 2002, 2006) who, although asked to learn 90 items in total, was given these in three sets of 30 items at a time. AK's results at 1 month on the -N+C set was 13/30, a significantly poorer result than CUB's 27/28 ($\chi^2 = 16.39$, $df = 1$, $p < .001$) while at 6 months she retained 9/30 compared to CUB's 23/28 ($\chi^2 = 11.13$, $df = 1$, $p < .001$). However, the exact influence of the number of items is hard to gauge since there are too few studies with enough variation in set size to enable clear conclusions to be drawn.

Therapy method and item selection

There was remarkable similarity in the way each participant was taught the correct phonology for the items they were learning, each being a version of picture-name paired learning. In Frattali's study (2004), if the participant was unable to name a picture, then he was given the correct phonology for the word (either by reading aloud or repeating) before learning. Three cases (Graham et al., 1999; Jokel et al., 2002; Snowden & Neary, 2002) used a "look, attempt to name, repeat after me/read aloud if unable to say" method which was also used in this study. Thus, each participant appeared to undergo remarkably similar treatment protocols.

There were differences in the way items were selected and how the items were supported within the therapy programme. In one of the experiments for example, DM (Graham et al., 1999) generated his own words using a category fluency paradigm, whereas in a later experiment and in the other studies, controlled, researcher-generated word lists were used. CR (Snowden & Neary, 2002) used self-study in a similar way to CUB but via a booklet rather than CD-ROM and links were made to her own environment. Both CR (Snowden & Neary, 2002) and AK (Jokel et al., 2002, 2006) made use of biographical information to expand the information provided during relearning. CR made explicit links between her word relearning and her environment, while AK provided her own definitions for the words in her learning set which were then used during relearning. CUB was able to relearn using a simplified therapy format, without reference to autobiographical information. When CUB did ask for assistance from her husband (for example to name the chair), she learned the word in the context of the phrase he provided for her, "table and chair", suggesting that she learned how to pair the picture to the phonology provided. Thus, it would appear that variations in therapy method and item selection do not account for CUB's therapy results.

Severity of semantic memory

Differences in severity have been suggested for the different patterns between DM (Graham et al., 1999, 2001) and CR (Snowden & Neary, 2002). DM's semantic impairment was mild while all the other cases showed moderate to severe semantic

impairment and anomia (see Table 1). If degree of semantic impairment and anomia were indicators of relearning ability, then CUB would be expected to perform in line with CR (Snowden & Neary, 2002), AK (Jokel et al., 2002, 2006) and Frattali's case (2004). Yet her relearning was superior and more resistant to loss across time than any of these. This is all the more perplexing given that, by the end of the study, CUB had the worst score on PPT (18/52).

Degree of atrophy

Table 1 sets out the scan reports for each case and from these it would be difficult to build a case for suggesting that differences in the location and extent of atrophy could account for the difference in relearning and retention. It may be that the relative sparing of other anatomical areas (such as the hippocampus and other medial temporal lobe structures) determines whether relearning can occur (assuming these structures underpin new learning: McClelland, McNaughton, & O'Reilly, 1995). However, reports based on visual inspection of the scans do not allow any formal exploration of any fine anatomical differences between the cases.

Premorbid experience and behavioural differences

There is little to suggest that premorbid differences could account for CUB's differential relearning. The occupations of KB and CK are not reported. However, DM was a retired surgeon, AK an arts organiser, and Frattali's (2004) case was a retired army officer. CUB was a high-functioning civil servant and so it would appear that all four were relatively well educated and motivated to retain and relearn. DM (Graham et al., 1999, 2001) and AK (Jokel et al., 2006) are both reported to have been fixated on the words they were losing, noting them down in a book, finding definitions in a dictionary, and devising their own ways to relearn them. CUB was similar to both of these in that she was obsessed by the loss of words and motivated to relearn them. It is interesting to note that the few therapy studies reported to date have been carried out with people who were fixated on their vocabulary loss. Consequently, there is a need to look across a more unselected group of people with semantic dementia to see whether others, who do not show this tendency, are also able to relearn.

Functional use/spontaneous speech

CUB's husband reported that, during and following therapy, she was able to use words she had learned at home appropriately. This functional use of therapy items in her spontaneous speech suggests that generalisation may have enabled a kind of "informal" practice, which allowed the words to be maintained for longer. This result is consistent with observations from other studies showing that everyday use and autobiographical knowledge enables longer-term retention of specific vocabulary as well as nonverbal activities such as object use (Bozeat et al., 2002; Snowden et al., 1994). The only aspect of her performance that does not seem to fit perfectly with this hypothesis is the (few) items that she named incorrectly at the 6-month follow-up (see Table 4). A number of these seem to relate more directly to everyday experience (bed, balloon, piano, hat, and cow) than some of the retained names (e.g., dromedary). Given this observation and the small number of unnamed items, future

studies are required that incorporate a deliberate manipulation and careful testing within the functional framework of the participant's life.

In summary, it would appear that generalisation to spontaneous speech may be the most likely cause of CUB's excellent learning and maintenance of learnt names despite no formal practice. Other factors may be important in the success of relearning in semantic dementia too, but these possibilities will need further exploration in future studies that allow direct comparisons across cases.

We finish with a brief note on the relearning mechanisms that are harnessed in these patients. In this regard the complementary learning systems (McClelland et al., 1995) are a useful framework in which to think about learning both for normal participants and patient groups. Using an implemented PDP model, McClelland et al. argued that learning is supported by two interactive mechanisms each with different characteristics. Hippocampal/medial temporal lobe structures allow for rapid learning of novel associations across modalities but do so at the cost of rigid representations that permit limited generalisation from one example to another. The counter-combination (slow learning that licenses generalisation) is afforded by neocortical structures including those in more lateral temporal lobe structures (which bear the brunt of the atrophy in semantic dementia). Learning in the normal, intact system involves the two learning mechanisms working in tandem, thereby allowing rapid initial representation of the experienced events/stimuli and then the gradual formation of representations that allow generalisation across examples. In patients with amnesia following medial temporal lobe damage, the rapid learning system is impaired but slow learning and subsequent generalisation are possible via the neocortical learning system.

Patients with semantic dementia can be considered as a reversal of this pattern; while they do have medial as well as inferolateral temporal lobe damage, the neural circuits underlying new learning have good metabolism (Nestor, Fryer, & Hodges, 2006)—thus allowing the rapid yet rigid medial temporal lobe system to function. In contrast, the inferolateral regions are often severely atrophied and have a corresponding hypometabolism (Nestor et al., 2006). This aligns clearly with the notion that these regions are involved in long-term semantic representations that allow for appropriate generalisations (Rogers et al., 2004). It is possible, therefore, that the picture name learning observed in CUB and other patients with semantic dementia primarily reflects the functioning of the medial temporal lobe system. In this context, one can view name learning as the novel association of a picture (visual representation) with a name (phonological representation). The medial temporal lobe system can rapidly learn to associate the two over a small number of learning trials but this learning is inevitably rigid in nature—thus the patients often reproduce the information in exactly the same way (e.g., preserving the learning order, cues and build-up information that was presented at the time of learning, etc.). With impoverished long-term semantic representations, the patients are then unable to generalise this learning appropriately and can only do so when the novel stimulus is very similar (visually for pictures or phonologically for words). Accordingly, the learning tends to be limited to the exact stimulus used in the learning trials and cannot be generalised to another example of the same type (e.g., CUB did not generalise the name “banana” from a standard picture to slices of banana, but did to visually similar depictions). Information encoded in medial temporal lobe structures needs to be actively maintained through continued re-exposure—i.e., by continuous, repeated deliberate practice or, as would appear to

be the most likely explanation for CUB's good retention, through generalisation to everyday use.

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APPENDIX

THERAPY AND CONTROL ITEMS WITH THEIR ENGLISH TRANSLATIONS

<i>Therapy set</i>	<i>Translation</i>	<i>Control set</i>	<i>Translation</i>
plátano	banana	ancla	anchor
cama	bed	flecha	arrow
campana	bell	cesta	basket
mariposa	butterfly	pájaro	bird
botón	button	hueso	bone
dromedario	dromedary	libro	book
vela	candle	cámara	camera
iglesia	church	queso	cheese
payaso	clown	cigarro	cigarette
peine	comb	reloj	watch
vaca	cow	corona	crown
tenedor	fork	perro	dog
guitarra	guitar	pato	duck
sombrero	hat	elefante	elephant
luna	moon	uva	grape
lápiz	pencil	corazón	heart
piano	piano	caballo	horse
tijeras	scissors	plancha	iron
zapato	shoe	cocina	kitchen
cuchara	spoon	ratón	mouse
sol	sun	nariz	nose
mesa	table	pera	pear
corbata	tie	pipa	pipe
hacha	axe	cochecito	pram
globo	balloon	radio	radio
bañera	bath	sandwich	sandwich
cinturón	belt	camisa	shirt
autobús	bus	paraguas	umbrella