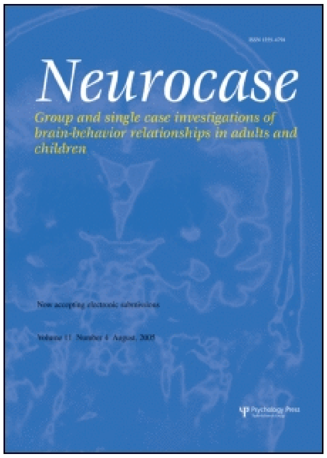

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The Natural History of Late-stage "Pure" Semantic Dementia

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Relatively little is known about the neuropsychological profile of late-stage semantic dementia. This article provides a detailed assessment of patient MK who, despite her very severe semantic impairments, remained cooperative to testing and, unusually, did not show additional behavioral/personality changes. Although MK's initial presentation was typical of semantic dementia (SD), her performance began to deviate from the normal pattern. She developed impairments of single word repetition and regular word reading, and began to produce phonological errors in picture naming and spontaneous speech. These deficits might suggest that late-stage SD includes an independent disorder of phonology. An alternative possibility, however, is that phonological processing cannot proceed normally in the face of profound semantic degradation. A series of experiments supported the latter explanation of MK's deficits. In picture naming, MK showed little effect of progressive phonological cueing, did not reveal an increased sensitivity to word length or phonological complexity and continued to show a high degree of item-specific consistency in both accuracy and errors: she tended to produce the same erroneous phonemes for each item. She remained sensitive to the effects of phonological similarity in immediate serial recall. Letter substitution errors in regular word reading were more common for lower frequency letters (e.g., Q, Z). These letters also produced more item errors in immediate serial recall, suggesting that a frequency-graded loss of letter knowledge, rather than separate orthographic and phonological deficits, accounted for the deficits in both of these tasks. These findings are discussed in terms of theories that posit strong interactivity between phonology and semantics.

Introduction

This article aims to provide a descriptive account of late-stage semantic dementia (SD). Although this condition has been the focus of extensive research for over a decade, relatively little is known about the neuropsychological profile of patients with very severe SD. It seems likely that this is because cases frequently become untestable at an earlier stage of the disease as a result of poor comprehension and/or behavioral disturbances. SD patients commonly develop behavioural and personality changes such as disinhibition, irritability, impulsivity and obsessions (e.g., patient MC; Hodges *et al*., 1992), presumably because the disease process increasingly affects basal frontal as well as temporal regions (Snowden *et al*., 1996). We report the evolution of cognitive impairment in one patient, MK, who remained unusually amenable to testing even when her semantic degeneration had become very severe, thus providing valuable information about the course of cognitive decline in SD.

In the early and middle stages of the disease, SD patients present with a highly uniform pattern of deficits and areas of preserved function; they show 1) a selective impairment of semantic memory causing severe anomia and comprehension difficulties, 2) sparing of other aspects of language, including phonology and syntax and 3) relative preservation of visual-spatial skills, non-verbal intelligence and day-to-day episodic memory (1989; Hodges *et al*., 1992, Snowden *et al*., 1996). This syndrome, which is associated with focal atrophy of the anterior inferior regions of the temporal lobe, is argued to result from specific degeneration of the semantic system. In line with this view, the spontaneous speech of SD patients is well articulated and grammatically correct but characterised by anomia and semantic errors (in particular, SD patients use high frequency, general words in the place of more specific names; e.g., "thing" for "biscuit"; Bird *et al*., 2000). In picture naming, SD patients make semantic errors (bear → "horse"), superordinate errors (bear → "animal") and, especially later on in the condition, produce uninformative descriptions (bear → "it's one of those things") and omissions (Hodges *et al*., 1995). Phonological cues usually have little effect on naming accuracy. Phonological errors occur very rarely and patients typically show excellent performance on single word repetition and phonological judgement tasks (Knott *et al*., 1997; Jefferies *et al*., 2005).

This circumscribed semantic impairment in SD has been shown to affect a variety of 'non-semantic' tasks (Patterson

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et al., in press), such as reading aloud (Hodges *et al*., 1992; Funnell, 1996; Jefferies *et al*., 2004b), immediate serial recall (Patterson *et al*., 1994; Knott *et al*., 1997; Jefferies *et al*., 2004a; Jefferies *et al*., 2005), lexical and object decision (Rogers *et al*., 2004) and the reproduction of line drawings after a brief delay (Bozeat *et al*., 2003). SD patients have poor reading of words with atypical spelling-sound correspondences that they no longer fully understand: they tend to pronounce these words as if they had regular correspondences (Funnell, 1996). They also have better immediate recall of strings of 'known' vs. semantically degraded items: the phonemes of poorly understood words are more likely to migrate and recombine with the phonemes of other list items, suggesting that word meaning helps to maintain short-term phonological memory (Patterson *et al*., 1994; Knott *et al*., 1997; Knott *et al*., 2000; Jefferies *et al*., 2004a; 2005). Severe semantic degradation in late-stage SD might be expected to produce similar difficulties on easier phonological tasks such as single word repetition.

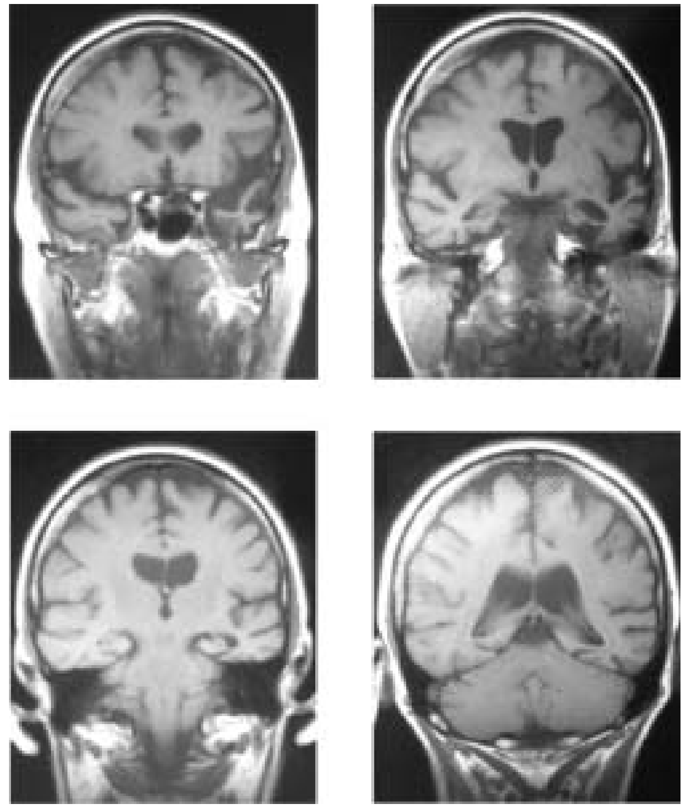

Fig. 1. MRI scan of patient MK from 2000:the four coronal sections show marked atrophy of anterior infero-temporal cortex.

Case Description

Initial Presentation

Patient MK, a right-handed British woman, had previously been employed in clerical work, having left school aged 17. In 2000, at the age of 66 years, she presented with a neuropsychological profile typical of SD. The main feature was a worsening difficulty in word finding and comprehension that her family had first noticed one or two years earlier. An MRI scan showed marked temporal lobe atrophy that was strongly lateralised to the left side (see Figure 1). Background neuropsychological scores are shown in Table 1. MK performed at or near floor on both verbal and visual semantic tests (e.g., the Pyramids and Palm trees test; Howard and Patterson, 1992). She was extremely anomic in picture naming and word fluency tasks. The majority of her naming errors were "don't know" responses (76%). She also made a small number of semantic errors (e.g., cow → "dog"), superordinate responses (e.g., owl → "bird") and descriptions (e.g., comb → "it's for your hair"). In contrast to her profound semantic impairments, she remained well oriented in time and place, although she was impaired on the Mini Mental State Examination (Folstein *et al*., 1975) because of her severe comprehension difficulties. Her speech was fluent, syntactically well formed and free from both phonological errors and articulatory problems. She had good autobiographical memory and episodic memory for recent events—she was able to recollect recent holidays and describe where she had lived previously. Her verbal short-term memory was normal as assessed by forwards and backwards digit span and she also performed well on the Corsi spatial span task (taken from Wechsler, 1997). In common with other SD patients, her immediate serial recall was more accurate for words that she still understood relatively well, compared with more semantically degraded words (Jefferies *et al*., 2004a, 2005). She showed a pattern of surface dyslexia in single word reading (e.g., she made regularisation errors, pronouncing PINT to rhyme with MINT) and surface dysgraphia in spelling tasks (e.g., writing TOMB as TOOM). She was able to copy the Rey complex figure accurately (taken from Lezak, 1976). She performed well on several visuospatial processing tasks from the VOSP (the Visual Object and Space Perception Test, Warrington and James, 1991), although she showed some weakness on several other tests from this battery. She was also impaired on the Progressive Matrices Test of non-verbal reasoning (Raven, 1962), which is unusual in mild to moderate SD. However, she did not show signs of disinhibition or other behavioral abnormalities and her family reported that her personality was unchanged.

Three to Five Years Post Onset

Over the next few years it seems likely that MK's semantic impairment became even more severe, although this is not well documented in Table 1 because her performance was already at floor when she was first tested. At around three years post onset, MK began to make phonological errors in spontaneous speech; these could be noted in normal conversation (e.g., she said "I wish I could file (find) it", while looking for something she had misplaced). At about the same time, MK started to make phonological errors in naming

Table 1. Neuropsychological Scores

Test	Max	MK				EK	Normal controls	
							M	SD
Years post onset		2	3	3.5	4	5		
Age		66	67	68	68	61		
MMSE[1]	30	21*	8*	4*	-	26	>24[a]	-
Naming	64	2*	3*	-	-	18*	62.3[b]	1.6[b]
Word-picture matching[†]	64	11*	11*	10*	-	39*	63.7[b]	0.5[b]
PPT: Pictures[5]	chance = 26/52	33*	31*	-	-	30*	51.1[b]	1.1[b]
PPT: Words[5]	chance = 26/52	26*	31*	-	-	35*	51.2[b]	1.4[b]
Category fluency (8 categories)	-	1*	1*	-	-	27	113.9[d]	12.3[d]
Letter Fluency (F, A, S)	-	2*	7*	7*	-	29	44.2[b]	11.2[b]
Coloured Progressive Matrices[2]	36	22*	15*	-	14*	33	-	-
Digit span: forwards[3]	-	5	5	5	4*	7	6.8[b]	0.9[b]
Digit span: backwards[3]	-	4	3	nu	-	4	4.7[b]	1.2[b]
Spatial span: forwards[4]	-	-	5	6	5	6	5 – 6[c]	-
Spatial span: backwards[4]		-	4	nu	-	4		-
Rey figure immediate copy[6]	36	30	35	31	31	36	34.0[d]	2.9[d]
VOSP: screening test	20	17	20	20	-	20		
VOSP: incomplete letters[7]	20	10*	4*	-	-	20	19.2[b]	0.8[b]
VOSP: dot counting[7]	10	10	10	10	-	9	9.9[b]	0.3[b]
VOSP: position discrimination[7]	20	17*	18	nu	-	20	19.8[b]	0.6[b]
VOSP: number location	10	6*	8	8	-	9	8.9	2.8
VOSP: cube analysis[7]	10	6	2*	-	-	10	9.7[b]	2.5[b]

Note.* denotes abnormal performance; nu denotes task not understood; span scores = maximum length repeated correctly; [†] word-picture matching task included nine semantically related distracters. [1]Mini-Mental State Examination (Folstein *et al*., 1975), [2]Raven's Coloured Progressive Matrices (Raven, 1962), [3]Weschler Memory Scale – Revised (Wechsler, 1987), [4]Weschler Memory Scale – III (Wechsler, 1997), [5]Pyramids and Palm Trees Test (Howard and Patterson, 1992), [6]Rey figure taken from Lezak (1976), [7]Visual Object and Space Perceptual Battery (Warrington and James, 1991). [a]Cutoff for normal performance, [b]Control data from Bozeat *et al*. (2000), [c]Normal range for age matched participants, [d]Control data from Hodges and Patterson (1995).

(e.g., fork → "fok", glass → "gas") and some of her responses bore even less resemblance to the target phonology (e.g., knife → "bull"). She produced nonwords in a letter fluency task (e.g., the letter A yielded the responses "dolly, atter, an, aner, afin, al, ando"). Her repetition was markedly impaired (toothbrush → "teabot"; close your eyes → "glose your eyes") and she began to make phonological errors in digit span (for example, the sequence "4, 2, 1, 6, 5" was repeated as "4, 2, light, side, fide"), producing the decline in digit span apparent in Table 1. She was only able to repeat 9/40 multisyllabic nonwords from the CN Rep test (Gathercole *et al*., 1994). On the minimal pairs task from the Psycholinguistic Assessment of Language Processing in Aphasia Test (PALPA; Kay *et al*., 1992), her accuracy was around 80% for both words and nonwords. MK's reading also deteriorated markedly in the same period and was increasingly dominated by implausible responses such as KEG → "ked". By five years post onset, MK's spontaneous verbal output consisted of a few ultra high frequency words and meaningless jargon. This is illustrated in the following excerpt, in which MK was talking about a cut on her finger.

> helluv-elev-uv-about about six or seven but it's terrible, yes". [*Examiner: Did you hurt it?*] "it's alright, it's alright but there, there-uv-there, there love there". [*When did it happen?*] "t . . . it . . . t . . . about ten elever there, elerveler there look, elerveler there look, cor yes.

MK's precipitous semantic deterioration also affected her performance on nonverbal tasks such as object decision; at three years post onset she was correct in 29/30 trials when the target had stereotypical features but at chance (12/30) when it had atypical features (task taken from Rogers *et al*., 2004). She also showed some unusual difficulties in delayed picture copying that seemed to stem from a failure to remember the identity of the object she was trying to draw (for example, when trying to reproduce a picture of a flower that she had seen moments before, she drew the petals correctly but then after a long pause, asked "is it a lady?" and added a human body underneath the flower head).

Despite these severe difficulties, MK was not globally impaired. There had been little deterioration in self-care, and she continued to clean the house and do the laundry. She remained sociable and retained her interest in gardening. Although she understood very little of what was said to her, she sometimes responded appropriately to stereotypical questions such as "how are you?" It appeared that her day-to-day memory was still relatively good: three years post onset she performed in the low average range on a test of face recognition (Wechsler, 1997). She could still find her way around very familiar places close to her home. She continued to use clocks and a calendar effectively until four years post onset. MK was still free from the behavioral characteristics of FTD. She remained cooperative in testing although her comprehension was so poor that she could typically only attempt tasks where the stimulus materials themselves indicated the required response (e.g., naming, reading and repetition). MK's performance on some tests of nonverbal reasoning and visual-spatial skills fell sharply, but these difficulties may have reflected failures to understand the instructions and possibly also semantic contributions to object recognition/segmentation—for example, she became severely impaired on the cube analysis subtest of the VOSP because she counted individual lines rather than the cubes they depicted.

Comment

Although MK's initial presentation was typical of SD, at a late stage her impairments began to deviate from the normal pattern. In particular, she began to make phonological errors in picture naming, spontaneous speech, single-word repetition and regular word reading. There are almost no published descriptions of late-stage SD presumably because formal testing typically becomes impossible. However, scant evidence suggests that such errors may emerge in late-stage SD quite commonly. Two patients in the Cambridge cohort that were followed over many years, PP (Hodges *et al*., 1992) and FM (Graham *et al*., 1995, 2001), began to make phonological errors in single word repetition, digit span, regular word reading and spontaneous speech at a late stage (unpublished observations). FM also made phonological errors in naming and word fluency tasks. In addition, Schwarz *et al*. (1998) outlined the evolution of SD in a single case over a period of 14 years. Initially this patient's impairments were confined to semantic memory. Eight years post-onset, he was uncooperative to testing but it was observed that his repetition was impaired.

MK's phonological errors, at least on the surface, are reminiscent of those observed in nonfluent progressive aphasia (NFPA; first described by Mesulam, 1982). NFPA is associated with atrophy affecting the left perisylvian cortex, centred on the anterior insula and frontal operculum (Nestor *et al*., 2003). Patients with NFPA frequently make phonological errors in spontaneous speech and picture naming (Croot *et al*., 1998; Watt *et al*., 1997) and they err on receptive phonological tasks (Croot *et al*., 1999), suggesting that they have a central phonological deficit. It has been argued that SD, NFPA and frontotemporal dementia (FTD) represent regional variations of a common pathology (or pathologies; Snowden *et al*., 1996; Hodges and Miller, 2001). Consequently, with disease progression, these distinct syndromes might be expected to develop common characteristics. There are some 'mixed' cases who show elements of both syndromes (Mesulam and Weintraub, 1992). For example, Papagno and Capitani (1998; 2001) described a patient with fluent speech who produced phonological errors sparing the initial phoneme and who later developed a severe semantic impairment. However, our observations suggest that SD may remain distinguishable from NFPA even at a very late stage of the disorder: speech in NFPA is slow/effortful and often ungrammatical: in contrast, MK's speech emerged effortlessly and remained prosodic and (as much as one can tell) syntactically correct even when it was characterised by jargon.

Although MK's phonological errors in naming, repetition and reading might point to an independent disorder of phonology, an alternative possibility is that phonological processing cannot proceed normally in the face of profound semantic degradation. As noted above, patients with mild to moderate SD show phonological errors in demanding tasks such as immediate serial recall and delayed repetition. These errors occur more frequently for semantically degraded items, suggesting that semantic memory makes an important contribution to phonological stability (Patterson *et al*., 1994; Knott *et al*., 1997, 2000; Jefferies *et al*., 2004a, 2005). By extension, extremely severe semantic deficits may result in phonological errors on less demanding tasks such as single-word repetition. Patient MK provided a unique opportunity to explore this hypothesis. We conducted a series of experiments to learn more about the origin of her phonological errors and their similarity with the errors observed in NFPA and mixed cases.

Experimental Investigations

Rationale

The following examination of MK's naming, repetition and reading three to four years post onset explored the nature and source of her atypical errors.

1. *Picture naming*: MK's picture naming performance was compared with that of a second SD patient, EK, who had much milder semantic impairment and did not make phonological errors in naming or spontaneous speech (see Table 1). We compared the influence of factors sensitive to phonological deficits in NFPA/stroke aphasia (word length and phonological complexity; Wilshire and McCarthy, 1993; Croot *et al*., 1998; Nickels and Howard, 2004) and factors sensitive to semantic impairment in SD (word frequency; Warrington, 1975; Lambon Ralph *et al*., 1998). MK's picture naming might be expected to show an increase in the influence of word length/phonological complexity if she had developed an additional impairment of phonology. We also examined test-retest consistency in

picture naming: this is typically high in SD, suggesting these patients have a semantic storage deficit (Coughlan and Warrington, 1981; Warrington and Cipolotti, 1996; Kertesz *et al*., 1998). MK might have shown reduced test-retest consistency if she developed a separable phonological impairment because phonological errors are often reported to have a probabilistic nature (Butterworth, 1992; see also Howard *et al*., 1984). Finally, we looked at the effect of progressive phonological cueing in picture naming. SD patients are typically insensitive to such cues, probably because their impoverished knowledge about the concept to be named provides so little activation of the appropriate phonological representation that the phonological cue is 'knocking on the door of an empty room' (Graham *et al*., 1995). If MK's unusual errors arose because of noise in the phonological system, however, then naming of items about which she still had some conceptual knowledge might benefit from phonological cueing.

2. *Reading*: we assessed the changing influence of regularity, word frequency and word length on MK's reading accuracy and errors longitudinally in order to better characterize her abnormal errors in this task. We also examined MK's knowledge of individual letters.
3. We compared picture naming, reading aloud and repetition for the same items. SD patients typically have profoundly impaired picture naming but good repetition and reading (at least for regular words). In contrast, patients with NFPA show more comparable difficulties across these tasks, in line with their phonological production problems (Croot *et al*., 1998). If MK had developed additional phonological deficits at a late stage of the disorder, one might expect to see a smaller difference in accuracy between naming and repetition/reading, relative to a milder SD patient, EK.
4. We examined the influence of phonological similarity in immediate serial recall. In healthy individuals, the poorer recall of phonologically similar items is usually taken as evidence for phonological coding in verbal short-term memory (Conrad, 1964; Conrad and Hull, 1964; Baddeley, 1966). Patients who have difficulty retaining phonological but not semantic information show a reduction in the size of this effect (Martin *et al*., 1994). The presence of a phonological similarity effect for MK would therefore suggest a relatively normal contribution of phonological processing to short-term memory.

Naming

Method

MK and EK attempted to name 212 pictures, including the 64 items referred to in Table 1 and an additional set of high frequency items. A subset of 88 items was re-presented a further three times to examine test-retest consistency. In order to maximize the number of phonological errors in the analysis, items were included if they had previously elicited a correct response or a phonological, semantic or superordinate error from MK, but not if she had responded with an omission or a description. It seems likely that MK's phonological errors would have been restricted to items that were at least partially semantically intact as no phonological activation would be expected for completely degraded items. EK's consistency on the 64-item set was also assessed over three test sessions. Finally, we presented progressive phonological cues for 25 pictures that frequently elicited phonological errors from MK in an attempt to maximize any potential impact of cueing. Cue length was increased one phoneme at a time until the entire target had been given.

Results

Accuracy was 11% for MK vs. 55% for EK. Both patients made omissions (MK: 30% of trials; EK: 18%), correct descriptions (MK: 23%; EK: 14%), semantic errors (e.g., horse → dog; 6% for both patients) and superordinate responses (horse → animal; MK: 2%, EK: 6%). However, MK showed additional types of error that EK did not make. Phonological errors occurred on 13% of trials (using a liberal criterion of one shared phoneme). Unrelated paraphasias (naming attempts that did not bear any phonological resemblance to the target) occurred on 8% of trials. Finally, 11% of MK's naming attempts were uninformative stereotypical phrases that she used repeatedly (e.g., "down through there"; "wandering around").

The factors affecting naming accuracy were analyzed with simultaneous logistic regression. The correlation between naming accuracy and word frequency (from Baayen *et al*., 1993) was significant for both patients but somewhat larger for EK (MK: $r = 0.25$, $p < 0.001$; EK: $r = 0.43$, $p < 0.0001$). The two patients displayed equivalent correlations between naming accuracy and phoneme length (both $r = -0.23$, $p < 0.001$). Neither patient showed a correlation between naming accuracy and an estimate of phonological complexity based on the number of phonemes per syllable. Naming success was predicted by patient identity (Wald = 75.90, $p < 0.0001$), word frequency (Wald = 14.38, $p < 0.001$) and phoneme length (Wald = 10.12, $p < 0.01$), but not phonological complexity (Wald = .03, n.s.). None of the interactions between these variables and patient identity significantly improved the fit of the model, suggesting that the impact of these factors did not differ for the two patients.

There were 83 phonological errors for MK; 58% contained at least one target phoneme whereas 42% shared at least half of the target phonology (including only the most accurate response from each trial). The majority (80%) contained the correct number of syllables. Those responses that did not preserve syllable length were typically shorter than the target (on 76% of trials). The syllable length of MK's responses was significantly associated with target length (C(N = 83) = .48, $p < 0.001$). MK's phonological errors also showed some sparing of first letter/sound knowledge: in CVC words (N = 29), accuracy was 69% for the first consonant, 38% for the vowel and 24% for the final consonant. In the complete set of phonological errors, 61% of initial consonants were produced correctly, compared with 34% of vowels and 50% of consonants in general. These findings fit with the observation that MK sometimes (on 4% of trials) indicated the correct starting letter of items she could not name. MK did not show a strong lexical bias in her phonological errors. 54% of close phonological approximations (that contained at least half of the target phonemes) were real words. This figure increased to 63% for more distant phonological errors (containing at least one target phoneme) and still further to 73% for naming attempts that did not share any of the target's phonology.

Table 2 shows the percentage of inconsistent and consistently correct/incorrect items for MK and EK on the high frequency set and, for EK only, on the 64-item set. Both patients showed a high degree of item-specific consistency across all of the adjacent test sessions for the high frequency set (MK: $C(N = 88) = .45 – .53$, $p < 0.0001$; EK: $C(N = 88) = .51 – .62$, $p < 0.0001$) and there was no evidence that the balance of consistent to inconsistent items was different for the two patients ($\chi^2(1) < 1$). However, EK's naming was substantially more accurate than MK's overall (72% vs. 21%). Table 2 also indicates consistency for the two patients when their total naming success was broadly equated (29% vs. 22%), by comparing EK's performance on the 64-item set with MK's performance on the high frequency set (only including data from the first three test sessions). Again, the proportion of consistent to inconsistent items was equivalent for the two patients ($\chi^2(1) < 1$). Simultaneous logistic regression indicated that MK's naming success on sessions 2, 3 and 4 was strongly predicted by her performance on the preceding test session, even when frequency and phoneme length were included in the model (Wald > 10.94, $p < 0.001$).

MK's phonological errors (defined as containing at least one target phoneme in the correct position) also showed a degree of item-specific consistency between test sessions (sessions 1 & 2: $C(N = 88) = .30$, $p < 0.01$; sessions 2 & 3: $C(N = 88) = 0.08$, n.s.; sessions 3 & 4: $C(N = 88) = .33$, $p < 0.001$). It was also noted that MK's erroneous phonology for each item tended to be consistent over time. For example, she consistently produced the /l/ in "glasses" as /r/; her responses were "greath, grease" (session 1), "grease" (session 2), "grass" (session 3) and "grease" (session 4). Although a general difficulty in producing the ends of words (see below) may have contributed to the commonalities between errors from different sessions, the same phonemes tended to intrude each time. A total of 43 phonological errors (preserving at least one phoneme in the correct position) occurred for items on multiple occasions, allowing the stability of the erroneous phonology to be examined. There were 71 intruded phonemes within these errors and 27 of these were repeated on the same item. Ten random rearrangements of the intrusions across items produced a mean of 9 repeated phonemes (SD = 4), indicating that this degree of consistency was very unlikely to occur by chance.

Table 2. Test-retest consistency in picture naming

Naming success	Same items (N = 88)		Difficulty matched items	
	MK	EK	MK (N = 88)	EK (N = 63)
0	64	15	66	67
1	14	13	15	8
2	7	7	6	10
3	8	9	14	16
4	8	57		

Note. Figures refer to percentage of items named 0/4–4/4 times.

Progressive phonemic cueing did not have a major impact on MK's naming accuracy even though cues were provided until the entire target had been given: accuracy was 5/25 without cueing and 8/25 with cueing.

Discussion

MK's phonological errors in picture naming typically preserved syllabic structure and initial phoneme. The sparing of the initial segments of words has been reported previously for NFPA (Croot *et al*., 1998) and for two cases with a mixture of fluent and nonfluent features (Papagno and Capitani, 1998, 2001; Patterson *et al*., Suzuki, Ijuin, & Tatsumi, 1998). However, other aspects of MK's naming responses did not resemble the pattern seen in NFPA. Whereas the naming success of patients with NFPA is strongly predicted by word length, reflecting the greater opportunity for error in words with more phonological segments, MK's naming was largely unaffected by length. She also continued to show a high degree of item-specific consistency in naming accuracy. This consistency extended to her phonological errors: she tended to produce the same erroneous phonemes each time an item was probed, suggesting that her errors were not the result of noisy phonological processing and instead reflected a distortion of phonological-lexical forms. Finally, in common with other SD patients, she did not show strong effects of phonological cueing in picture naming even for a set of items that typically gave rise to phonological errors.

Reading Aloud

Method

MK was asked to read 252 monosyllabic words (minimum length = 3 letters, maximum = 6) that crossed regularity of spelling-sound correspondences (e.g., regular words such as "mint" vs. irregular words such as "pint") and word frequency (high, medium and low frequency words; list from Patterson and Hodges, 1992). MK read these words at two, three and four years post onset.

Results

Figure 2 indicates accuracy of reading aloud as a function of regularity, word frequency and time post onset. At initial testing,

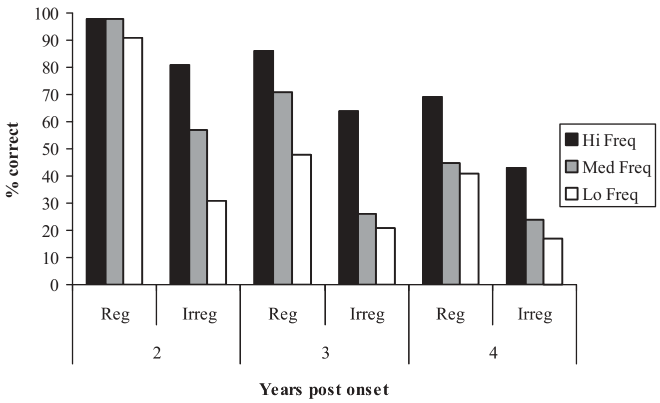

Fig. 2. MK's reading performance as a function of regularity, word frequency and years post onset, N = 252 divided equally between the conditions. Data include first response only.

Table 3. Length effects and errors in reading aloud as a function of years post onset

	Years post onset	2	3	4
	Age	66	67	68
Accuracy	3–4 letters	85	66	54
(% correct)	5–6 letters	75	48	32
Errors	LARC (plausible)	73.8	45.4	28.9
(% total errors)	Mixed	6.6	17.6	27.0
	Other (implausible)	19.7	37.0	44.1

Note. N = 252. Figures are percentage of total errors, collapsing across the regularity by frequency conditions. Only the first response was included for each trial. LARC = legitimate alternative reading of components.

MK's reading pattern was one of classic acquired surface dyslexia, that is, a massive advantage for words with regular spelling-sound correspondences, with reading success for the irregular words (but not the regular ones) strongly modulated by frequency. At subsequent testing rounds, however, although the advantage for regular > irregular words was maintained, frequency had a major impact on both word types. Simultaneous logistic regression showed independent effects of four variables on MK's reading performance: time post onset (Wald = 22.58, $p < 0.0001$), word frequency (Wald = 35.09, $p < 0.0001$), regularity (Wald = 25.83, $p < 0.0001$) and number of letters (Wald = 7.65, $p < 0.01$). The interaction between time post onset and regularity significantly improved the fit of the logistic regression model, showing that the regularity effect diminished as the condition progressed (Wald = 4.65, $p < 0.05$).

MK's errors are shown in Table 3. Errors were classified as 'LARC' (Legitimate Alternative Reading of Components: Patterson *et al*., 1995) if they were plausible in the sense that each component of the word was pronounced in a way that would be appropriate in a different item. LARC errors occurred for regular words (HOOT pronounced to rhyme with 'foot') as well as irregular words (BLOOD pronounced to rhyme with "good"). Strict regularizations (BLOOD pronounced to rhyme with "mood") formed a subset of these errors. 'Other' errors were implausible pronunciations of the letter string. Many of these errors apparently resulted from letter misidentifications; examples are KEG → 'ked' and SOB → 'sop'. 'Other' errors could also result from the addition or deletion of letters/sounds (TRUMP → 'strump'). Finally, 'mixed' errors contained both plausible and implausible elements, as in HOOT → "hoad". MK showed a sharp increase in implausible pronunciations over the three testing sessions, significantly changing the balance of implausible 'other' to LARC/mixed errors ($\chi^2(2) = 11.1$; $p = .004$).

We examined the letter substitution errors in greater detail. Only those errors in which a single consonant had clearly been misread as another single consonant were included (e.g., BULL → "full"). Multi-letter substitutions were excluded (GLIDE → "bride"). In order to facilitate calculation of the rate at which each substitution was expected by chance, the analysis was confined to the initial letter of each word. Errors were pooled across the three testing sessions (N = 2, N = 29 and N = 52 on the first, second and third sessions respectively). Letter misidentifications occurred more frequently for low frequency letters such as Q, W and Z. There was a negative correlation between letter frequency (data from Baddeley *et al*., 1960) and the number of substitutions ($r = -.46$, one-tailed $p = .04$). In addition, MK showed a significant level of consistency in her substitutions, with 70% of substitutions involving letter pairs that had already been exchanged at least once. The most frequent substitution was p → b, followed by m → b. In order to establish an appropriate baseline level of consistency, the letters were pseudo-randomly re-paired (in such a way that the letters within a pair were not identical) and the number of repeated pairs was counted (M = 39.4%; SD = 2.28). The observed number of repetitions exceeded the number expected by chance. MK's substitutions did not, on the whole, reflect global visual similarity (as measured by the extent to which normal subjects mistake one letter for another when reading from a long distance or in eccentric vision: data from Bouma, 1971). The average percentage of trials in which Bouma's participants confused the pairs of letters exchanged by MK was 1.63. This can be compared with the same measure for ten pseudo-random rearrangements of the letters (M = 1.54, SD = 0.25). The observed figure did not exceed the value expected by chance.

Single Letter Tasks

Method

MK was asked to (1) name 52 letters in a random order (the full alphabet in upper and lower case); (2) perform spoken-written letter matching with the full alphabet; (3) make same/different judgements for letters in upper and lower case; (4) write the letters of the alphabet to dictation in a random order.

Results

(1) MK named 94% of upper case and 92% of lower case letters (averaging across two presentations) at three years post onset. At 4.5 years post onset, accuracy had dropped to 85% and 73% respectively. Her errors involved both the production of incorrect letter names (y → "J") and non-letter responses (q → "kwee"; h → "hay"). Many of her errors involved low frequency letters (e.g., Q, Z). The letters that she read correctly were significantly more frequent than the letters that she read incorrectly (using estimates of letter frequency from Baddeley *et al*., (1960); $t(258) = 2.67$, $p < 0.01$). (2) MK was also impaired at spoken-written letter matching (85% of upper case and 81% of lower case letters correct) at three years post onset. (3) On cross-case matching, MK scored 91/96 both times she was tested at three and four years post onset. (4) MK successfully wrote 50% and 46% of letters to dictation at three and four years post onset. Her errors were a combination of incorrect letters (V → Z; W → G) and multi-letter responses that spelt out the letter name (Q → cue; H → ach). The letters that she wrote correctly were significantly more frequent than the letters she failed to write ($t(76) = 5.74$, $p < 0.0001$).

Discussion

Implausible errors in regular word reading developed at about the same time as phonological errors in naming, spontaneous speech and repetition. This produced a breakdown in the surface dyslexic pattern which is typically observed in SD. MK made substitutions of one letter for another during word reading and also had poor letter recognition, although this apparently did not result from confusions between visually similar letters but instead reflected a frequency-dependent loss of knowledge about letter shapes. Semantic degradation in SD is highly sensitive to word frequency (Funnell, 1995) and it seems likely that the same might hold for individual letters at a later stage of the disease.

Degraded knowledge of single letters and the emergence of implausible reading responses have been observed in another late-stage SD patient, FM (Graham *et al*., 1997, 2001). FM developed difficulties in cross-case letter transcription and letter naming tasks. These problems co-occurred with the production of phonologically implausible responses in reading and spelling which became increasingly dominant: at a late stage, FM's written output was primarily jargon that bore little resemblance to the target words. FM's spelling did, however, continue to obey orthotactic rules: she produced few illegal letter combinations. She was also sensitive to letter frequency and eventually produced only high frequency letters. Graham *et al*. (2001) suggested that FM's jargon dysgraphia might have arisen from her very severe semantic impairment rather than an additional peripheral impairment because she continued to show effects of semantic knowledge in spelling (i.e., her responses were closer to the target for better known than for very degraded words). In late-stage SD, orthographic and phonological outputs show similarities: jargon spelling shows a weakened influence of semantics/phonology on orthographic responses, whereas reading aloud shows a loosening of semantic and orthographic constraints on phonological responses. The suggestion that these effects result from severe semantic breakdown which impinges on the knowledge of individual letters and their corresponding sounds in a frequency-sensitive way is more parsimonious than positing separate deficits of semantics, orthography and phonology in late-stage SD.

Naming, Reading and Repetition of the Same Items

Method

MK and EK performed naming, reading and repetition tasks with the items used to assess picture naming consistency, and also with the 64-item set (total N = 145; six items were excluded either because they appeared in both word sets or because they were correctly named but by an alternative word not included in the repetition/reading tests: e.g., "settee" for sofa). The patients attempted to repeat each item both immediately and after a five-second delay filled by the patient and experimenter counting aloud from one to ten. A subset of items was selected to examine the effect of word length in each task (N = 74). Short (monosyllabic) and long items (two to four syllables) were matched on an item-by-item basis for word frequency.

Results

Table 4 shows the percentage of items that were correct in each task, and gives an indication of the types of errors that occurred. MK showed significant consistency between naming and repetition/reading (see Table 5). Accuracy (in terms of both number of items and average percentage of phonemes correct) followed the pattern immediate repetition = reading > delayed repetition >> naming. The difference between immediate repetition (mean = 94% of phonemes correct, SD = 13.3) and reading (91% of phonemes correct, SD = 14.1) did not reach significance ($t(144) = 1.81$, $p = 0.07$). Reading was more accurate than delayed repetition (82% of phonemes correct, SD = 26.2; $t(144) = 3.74$, $p < 0.001$). There was a considerable advantage for delayed repetition over naming (18% of phonemes correct, SD = 32.9; $t(144) = 18.34$, $p < 0.0001$). EK showed a similar pattern across these tasks; accuracy was highest for immediate repetition and lowest for picture naming, although reading and delayed repetition did not significantly differ. The difference between delayed repetition and naming was equivalent for MK and EK (43% vs. 46%). Similarly, MK did not show a reduced difference between reading and naming (59% for MK vs. 39% for EK).

MK's accuracy in reading was not influenced by syllable length (78% and 76% correct for short and long words respectively). For immediate repetition, the difference between short and long words approached significance (92% vs. 73%, Fisher's Exact Test, two-tailed, $p = .06$). Delayed repetition was significantly poorer for longer words (78% vs. 43%, Fisher's Exact Test, two-tailed, $p = .004$). Naming was at floor for both long and short words.

Table 4. Naming, reading and repetition of the same items

	MK				EK			
	Naming	Imm rep	Del rep	Reading	Naming	Imm rep	Del rep	Reading
Correct	10.3	79.3	53.1	69.0	49.0	100	95.2	88.3
Semantic/superordinate/ descriptive errors	29.7	0	0	0	31.0	0	0	0
Phonological errors total	15.1	20.7	43.4	31.0	0	0	2.1	11.7
Close (≥50% shared phonemes)	4.8	18.6	38.6	31.0	0	0	2.1	11.7
Distant (≥1 shared phoneme)	10.3	2.1	4.8	0	0	0	0	0
Unrelated errors	13.1	0	2.1	0	0	0	0	0
Omissions	13.1	0	1.4	0	19.3	0	0	0
Other errors	18.6	0	0	0	0.7	0	2.8	0

Note. N = 145. Errors expressed as a percentage of trials presented. Imm/del repetition = immediate/delayed repetition.

Table 5. Item-specific consistency between naming, repetition and reading

		Consistency in accuracy (N = 145)						% phonemes correct	
	Task	1,1	1,0	0,1	0,0	C	*p*	Correlation	*p*
MK	Naming & immediate repetition	10.3	0.0	69.0	20.7	0.17	<0.05	0.18	<0.05
	Naming & delayed repetition	6.2	4.1	46.9	42.8	0.05	n.s.	0.02	n.s.
	Naming & reading	9.7	0.7	59.3	30.3	0.18	<0.05	0.18	<0.05
	Immediate repetition & reading	59.3	20.0	9.7	11.0	0.24	<0.01	0.11	n.s.
	Delayed repetition & reading	38.6	14.5	30.3	16.6	0.09	n.s.	0.03	n.s.
	Immediate & delayed repetition	53.1	26.2	0.0	20.7	0.48	<0.001	0.45	<0.0001
EK	Naming & reading	44.1	4.1	44.1	7.6	0.09	n.s.	0.05	n.s.

Note. Figures show percentage of items correct in both tasks (1,1), one task only (1,0 or 1,0), and neither task (0,0), and correlations between the percentages of phonemes produced correctly in the pairs of tasks. EK's scores for immediate and delayed repetition are omitted because her performance approached ceiling.

Discussion

MK's substantially lower accuracy for naming than reading and repetition concords with the view that her single-word production was dominated by semantic impairment. She was extremely poor at generating phonology from semantics but showed considerably improved performance when phonology was specified by the task (repetition) or constrained by orthography (reading aloud), in line with other SD patients. Naming, reading and repetition have also been compared in two patients with NFPA; they showed more similar levels of performance on the three tasks, in line with their phonological production problems, and were somewhat less accurate in immediate repetition than reading, in contrast to MK (Croot *et al*., 1998). MK showed effects of syllable length in delayed repetition but not reading, and continued to show a considerable advantage for reading/repetition over naming for longer items. In contrast, the NFPA patients studied by Croot *et al*. were highly sensitive to length in every task and showed more equivalent accuracy across the tasks for longer words. Consequently, we did not obtain positive support for the view that MK's impairments overlapped with features of NFPA in the advanced stages of the disease. Instead, MK's item-by-item consistency across naming, reading and repetition coheres with the proposal that her errors in these tasks were a consequence of her severely degraded semantic representations—this would be anticipated if word meaning plays an important role in constraining phonological production in repetition and reading.

Phonological Similarity in Immediate Serial Recall

Method

Four years post-onset, we examined the effect of phonological similarity in immediate serial recall with higher frequency letters (E, C, D, T, vs. A, I, L, O) and lower frequency letters (B, G, P, V vs. F, K, X, Y). Each list of four items was read aloud at a rate of one item per second. There were 20 lists in each condition, presented in an alternating fashion. Three measures were examined: 1) items recalled in the correct position, 2) order errors (items recalled in the incorrect serial position) and 3) item identity errors (items not presented anywhere in the list: this category included intrusions, repetitions, phonological errors and occasional omissions).

Results

The data are shown in Table 6. We avoided confounding differences in order memory with item recall accuracy by dividing the number of order errors in each list by the total number of items recalled correctly in any order (Table 6 shows order errors as a percentage of items presented). Factors affecting item and order errors were analyzed using ANOVA. There was a significant main effect of phonological similarity ($F(1,76) = 4.70$, $p < 0.05$) and a three-way interaction between phonological similarity, letter frequency and error type ($F(1,76) = 4.54$, $p < 0.05$). Phonological similarity influenced the incidence of order errors ($t(78) = 2.38$, $p < 0.05$) but not item errors ($t(78) <1$), whereas letter frequency impinged on item errors ($t(78) = 2.77$, $p < 0.01$) but not order errors ($t(78) <1$).

Discussion

MK had poorer immediate serial recall for phonologically similar letters when she was first tested two years post-onset (Jefferies *et al*., 2004c). She still showed this effect of phonological similarity on immediate serial recall at a late stage of the condition when the frequency of the phonologically similar/dissimilar letters was equated. Phonologically similar letters were more likely to be recalled in the wrong order, indicating that MK continued to rely on a phonological code to maintain sequences of items. She also showed an effect of letter frequency on recall: item errors occurred at a higher rate for low frequency letters such as X and V, that she was less able to name, read and recognize. Similarly, in mild-moderate SD, well-known words are recalled more accurately than semantically degraded words and semantic status affects the likelihood of phonological/item errors but not order errors (Jefferies *et al*., 2004a). The fact that letter frequency effects were observed in verbal short-term memory as well as orthographic tasks supports the view that MK's non-plausible letter substitution errors in reading aloud resulted from a frequency-modulated loss of letter knowledge rather than a separate orthographic deficit.

General Discussion

We examined a patient with late-stage semantic dementia, MK, who was a relatively 'pure' case in the sense that she did not develop the behavioral symptoms of fronto-temporal dementia. She remained unusually cooperative to testing, yielding useful information about the evolution of this condition. When her semantic impairments became very severe, MK started to make phonological errors in spontaneous speech, regular word reading, picture naming and immediate single-word repetition. SD cases with mild to moderate semantic impairments do not typically produce such errors (e.g., Hodges *et al*., 1992; Snowden *et al*., 1996). We attempted to ascertain whether MK's unusual errors resulted from a separable phonological problem that developed alongside her primary semantic impairment. Wide-ranging deficits may emerge in late-stage SD as the disease spreads to new brain areas; MK's phonological errors might have reflected this progression. However, an alternative interpretation is that phonological processing cannot proceed normally in the face of profound semantic degradation.

A series of experiments revealed a surprising paucity of evidence for a separable phonological impairment. MK's picture naming remained highly consistent across test sessions and did not show an increase in the impact of a phonological variable, word length, relative to the performance of a second patient who had a milder semantic impairment and deficits more typical of SD. MK's phonological errors typically affected the same items on each test session and, strikingly, there was even consistency in the phonemes that intruded into each item. In contrast, phonological errors that arise from noise should be more frequent for longer words, which present a greater opportunity for error (see Croot *et al*., 1998) and are expected to have a probabilistic nature reducing item-specific consistency (Butterworth, 1985, 1992). These findings are therefore incompatible with the view that the errors resulted from noise in the phonological system and are perhaps indicative of a distortion of the phonological space itself (see below). MK continued to show substantially better reading/repetition than picture

Table 6. Phonological similarity and letter frequency in immediate serial recall

		Correct in position		Order errors		Item errors	
Frequency	Phonological similarity	M	SD	M	SD	M	SD
High	Dissimilar	57.5	25.8	15.0	22.1	27.5	13.8
High	Similar	50.0	25.6	16.3	18.6	33.8	14.7
Low	Dissimilar	51.3	20.6	6.3	13.8	42.5	16.4
Low	Similar	36.3	27.5	26.3	28.6	37.5	15.2
High	Average	53.8		15.6		30.6	
Low	Average	43.8		16.3		40.0	
Average	Dissimilar	54.4		10.6		35.0	
Average	Similar	43.1		21.3		35.6	

Note. Figures denote percentage of items presented.

naming, indicating that her single word production was still dominated by her severe semantic deficits. This advantage for repetition/reading over naming was not reduced relative to a second patient with milder SD. She also showed strong item-specific associations between naming and repetition/reading, in line with the view that her central semantic impairment underpinned her poor scores in these apparently "non-semantic" tasks. If MK had developed significant phonological and orthographic problems in addition to her semantic deficits, this consistency might have been reduced. Finally, MK continued to show a significant effect of phonological similarity in immediate serial recall when the phonologically similar and dissimilar items were matched for frequency. This effect is typically ascribed to the use of a phonological code in verbal STM (Conrad, 1964; Conrad and Hull, 1964; Baddeley, 1966), and might be eroded in patients with a primary phonological deficit.

In summary, MK's performance across a range of tasks was dominated by semantic degradation; we found little evidence of an additional phonological deficit. Although MK showed impairments of single word repetition and regular word reading, these appeared to be strongly related to her semantic difficulties. MK's failure to repeat words that she did not understand can therefore be viewed as an extension of the well-documented repetition impairment shown by milder SD patients for semantically degraded items. Several studies have demonstrated that demanding phonological tasks such as immediate serial recall and delayed repetition are disrupted in SD, and recall under these conditions is typically poorer for words that are more semantically degraded, consistent with the view that semantics makes a crucial contribution to phonological processing (Patterson *et al.*, 1994; Knott *et al.*, 1997, 2000; Jefferies *et al.*, 2004a, 2005). In patients with mild to moderate SD, these effects may only be observed with greater phonological loads and at longer time delays, but by extension, very severe semantic degradation might be expected to disrupt less challenging tasks like single immediate word repetition. The worsening phonological problems in late-stage SD therefore cohere with the suggestion that semantics plays a major role in phonological processing.

Semantic input to the phonological system may help to maintain phonological activation as it decays (Martin and Saffran, 1997). In addition, semantics can usefully constrain phonological activation because every time a particular word is spoken or heard-and-comprehended, specific semantic and phonological patterns co-occur for that word (Patterson *et al.*, 1994). According to some theoretical standpoints, distortion of the phonological space is a predictable consequence of severe semantic degradation because phonological representations are acquired in the presence of semantics. For example, in the "model-T" framework of Plaut and Kello (1999), activation between acoustic, phonetic and semantic representations is accomplished through a common set of hidden units, which in effect form a "phonological" space. Given the high degree of systematicity between acoustic and phonetic representations, the representational space will predominantly reflect these associations. In addition, however, the model is required to transform these intermediate representations into meaning, and thus semantic memory also influences the formation of this "phonological" space to at least some degree. As a result, semantic degradation will disrupt phonological processing. This functional impairment of phonology should eventually affect all tasks involving spoken output, including spontaneous speech, picture naming, reading and repetition. Indeed, MK's phonological errors in naming were non-random across trials, suggesting that her phonological-lexical representations may have become distorted. By the end of the study, her spontaneous verbal output contained fluent jargon; however, phonemic jargon can preserve the basic phonological and prosodic properties of the language in the absence of any recognizable lexical forms (Hanlon and Edmondson, 1996).

There are some important differences between naming, reading and repetition, however, which are likely to influence the way semantic degradation impairs performance. In repetition/reading, the spoken or written stimulus provides a model that strongly drives phonological output; consequently, in these tasks, SD patients make phonological errors and very few omissions (see Table 3). In picture naming, where there is no phonological stimulus, omissions occur more commonly, perhaps because under conditions of severe semantic degradation, it is unlikely that a single candidate response at the phonological level will be sufficiently activated to exceed a criterion for speech production. Disease progression is unlikely to produce a lessening of this problem, so why do phonological errors only emerge in spontaneous speech/picture naming at a late-stage of the condition? There are at least two possibilities. First, MK's criterion for responding in picture naming may have changed: she may have been increasingly willing to produce an incorrect response at a late-stage, perhaps because she developed severe difficulties in recognizing nonwords in her own output. Graham *et al.* (2001) argued for a change in response criterion for the late-stage SD patient FM: at one stage, FM produced jargon responses in spelling to dictation but omission errors in written picture naming; a year later, she could be induced to make jargon responses in picture naming as well. An alternative possibility is that the presence of a phonological model in repetition/reading encourages a lower response criterion relative to picture naming. We did not observe strong evidence that a change in response criterion led to the appearance of phonological errors in MK's picture naming. Closer inspection of the 64-set longitudinally reveals that she did not attempt to produce the phonology of the majority of items she had previously omitted. Although she made more omission errors when she was first tested at two years post onset (73% of trials) than at three or four years post onset (50% and 19% of trials respectively), this decrease in omissions was linked to an increase in vague descriptions (e.g., TORTOISE → "he wanders around": 6%, 34% and 50% of trials at two, three and four years post onset), rather than an increase in phonological errors (2%, 6% and 9% of trials). Instead, MK's phonological errors were confined to very high frequency items that may have remained at least partly understood (and therefore able to drive some phonological activation). The finding that MK made phonological errors consistently on the same items also concurs with the view that these items were more semantically intact.

We have suggested that MK's phonological errors may reflect the impact of severe semantic impairment on phonological processing in a highly interactive system. According to this account, phonological errors should be the norm in late-stage SD. In contrast, if such errors arise because of an independent impairment of phonology, not all late-stage SD patients will necessarily develop them. It is difficult to establish the generality of our findings with MK because few patients with initially prototypical SD have been followed to a late stage. It is however useful to note that two SD patients who were followed over a long period developed some of the characteristics discussed here. Both PP (Hodges *et al*., 1992) and FM (Graham *et al*., 1995; Graham *et al*., 2001) began to make phonological errors in single word repetition, digit span, regular word reading and spontaneous speech. FM was also observed to make phonological errors in naming and word fluency tasks. Word repetition after a brief filled delay became impossible for FM (she produced the name of a familiar shop, "Sainsbury's", on every trial, no matter what the target word). FM's speech at this stage was heavily reliant on stereotyped utterances: she used the phrase "special place" in a strikingly similar way to MK's expressions "down through there" and "wandering around". These parallels between the late-stage deficits of MK and two other patients suggest that the pattern we have documented here may be relatively widespread.

MK, PP and FM all had bilateral but asymmetrical temporal lobe atrophy that was more severe on the left side. Although it will be necessary to follow a larger number of cases to confirm this suggestion, patients with predominantly left-sided atrophy may be more likely to develop the late-stage pattern of neuropsychology described here. There are two potential reasons for this predicted influence of laterality. Previous reports indicate that patients with more right-than left-sided atrophy (R>L) show greater behavioral and emotional changes than patients with predominantly left-sided atrophy (Edwards Lee *et al*., 1997; Perry *et al*., 2001; Rosen *et al*., 2002; Thompson *et al*., 2003). These behavioral impairments often make detailed assessment impractical and so R>L patients may become untestable before their semantic deficits are severe enough to produce problems in phonological processing for single words. An alternative (or additional) proposal arises from a neuroanatomically-constrained model of semantic memory and speech production (Lambon Ralph *et al*., 2001). In this model, concepts are represented bilaterally across left and right temporal regions whilst left hemisphere areas support phonology. This means that the integrity of the left hemisphere semantic region is much more critical in speech production (which requires interaction between semantic and phonological representations). The late-stage pattern exhibited by MK, PP and FM may reflect an extrapolation of this model—the exaggerated phonological distortion may result from extremely severe damage to the left hemisphere portion of the semantic system.

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