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Deep Dysphasic Performance in Non-fluent Progressive Aphasia: a Case Study

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Abstract

We present a patient (PW) with non-fluent progressive aphasia, characterized by severe word finding difficulties and frequent phonemic paraphasias in spontaneous speech. It has been suggested that such patients have insufficient access to phonological information for output and cannot construct the appropriate sequence of selected phonemes for articulation. Consistent with such a proposal, we found that PW was impaired on a variety of verbal tasks that demand access to phonological representations (reading, repetition, confrontational naming and rhyme judgement); she also demonstrated poor performance on syntactic and grammatical processing tasks. However, examination of PW's repetition performance also revealed that she made semantic paraphasias and that her performance was influenced by imageability and lexical status. Her auditory–verbal short-term memory was also severely compromised. These features are consistent with 'deep dysphasia', a disorder reported in patients suffering from stroke or cerebrovascular accident, and rarely reported in the context of non-fluent progressive aphasia. PW's pattern of performance is evaluated in terms of current models of both non-fluent progressive aphasia and deep dysphasia.

Introduction

In this paper we examine the language performance of PW, diagnosed with non-fluent primary progressive aphasia. Primary progressive aphasia is characterized by a progressive degeneration in speech production over at least a 2-year period, with no evidence of generalized dementia, otherwise normal activities of daily living, and relatively spared non-verbal abilities as determined by neuropsychological testing (Mesulam, 1982, 1987). Primary progressive aphasia typically appears in the sixth decade of life, but has been seen across the age range 45–70 years and has a mean duration of approximately 8 years (Snowden *et al.*, 1996). Over the last decade it has been determined that such patients can be further subclassified into two major types: fluent and non-fluent progressive aphasia.

Fluent progressive aphasia is characterized by syntactically and grammatically normal spontaneous speech that is typically 'empty' of content (Hodges *et al.*, 1992). Such patients also become profoundly anomic as their condition worsens, and display a similar degeneration on tests of comprehension and semantic memory (Poeck and Luzatti, 1988; Breedin *et al.*, 1994). To reflect the central semantic deficit underlying this degeneration, several authors have preferred the term 'semantic dementia' (Hodges *et al.*, 1992; Breedin *et al.*, 1994; Graham *et al.*, 1995; Snowden *et al.*, 1996).

Non-fluent progressive aphasia is characterized by agrammatic and stuttering spontaneous speech, which often contains phonemic paraphasias. However, despite impaired syntactic and grammatical processing, performance on comprehension of single words is typically spared. Normal performance is often seen on tests of object identification, spatial ability, non-verbal ability, episodic memory and calculation (Snowden *et al.*, 1996). Although a large number of case studies of non-fluent-type progressive aphasia have been published, the majority of such reports have been clinical in nature (e.g. Kartsounis *et al.*, 1991; McDaniel *et al.*, 1991; Caseli and Jack, 1992; Thompson *et al.*, 1997) and few have attempted to conduct a detailed examination of language breakdown in such patients. For this reason, our understanding of the nature of language impairment in non-fluent progressive aphasia is somewhat limited. Croot *et al.* (1998, 1999) have, however, recently provided two extensive studies in an attempt to fill the theoretical vacuum.

Croot *et al.* (1998) report a systematic study of language performance in two patients, PG and LM, diagnosed with non-fluent progressive aphasia. They found that both patients demonstrated a significantly higher rate of phonologically correct responses in single word reading and repetition tasks, relative to picture naming. Overall, they concluded that both patients were impaired in phonological processing and that differences in severity across tasks occurred because phono-

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logical representations are more directly specified in reading and repetition tasks than in naming. In a subsequent study, Croot *et al.* (1999) described brothers diagnosed with non-fluent progressive aphasia. Although initially CB and RB showed a pattern of poorer performance at naming tasks relative to repetition and reading (consistent with PG and LM), the brothers did not show the same decline in performance across all three tasks. In this case, RB demonstrated consistently better performance at repetition relative to reading. However, the reverse was true of CB, who performed better on reading. Both patients remained poorer at confrontational naming relative to both reading and repetition. Closer examination of CB's repetition performance revealed an effect of imageability, which the authors suggest implies that the patient was becoming increasingly reliant on semantic processing when repeating single words. Croot *et al.* (1999) concluded that although superficially both patients were severely impaired in terms of language production, careful examination had determined differences in terms of the presence of particular language function deficits.

Croot *et al.* (1999) provide evidence that performance of non-fluent progressive aphasics on repetition tasks can be quite variable. A recent review of 112 patients with non-fluent progressive aphasia determined that 10% of non-fluent progressive aphasics demonstrate a severe repetition deficit by the third year of symptom onset, and this figure increased to 20% by the ninth year from onset (Westbury and Bub, 1997). Since this study there have been two reports of severe repetition difficulties in patients with non-fluent progressive aphasia (CB: Croot *et al.*, 1999; CO: Majerus *et al.,* 2001). In both cases these patients showed better repetition of high relative to low imageability words and made semantic paraphasias. Interestingly, this aspect of their profile is similar to that seen in patients with 'deep dysphasia'.

Deep dysphasia was first described by Michel and Andreewsky (1983), and is a disorder of repetition that is characterized by: (1) semantic paraphasias in repetition, (2) heavily impaired non-word repetition, (3) significantly better repetition of high relative to low imageability words and (4) severely impaired auditory–verbal short-term memory. Since this seminal paper, subsequent reports of patients with deep dysphasia have been rare, with less than 20 being published (e.g. Marshall and Newcombe, 1988; Katz and Goodglass, 1990; Coslett, 1991; Martin and Saffran, 1992; Butterworth and Warrington, 1995). There is also some debate as to the unitary nature of this syndrome [cf. Trojano and Grossi (1997) and the subsequent reply by Martin (1997)]. However, several interesting additional impairments have been established from the small group of patients who have been described. Semantic paraphasias in writing to dictation have also been reported and reading performance can be surface dyslexic (e.g. Howard and Franklin, 1988). Deep dysphasic patients have also been reported with severe auditory comprehension problems (e.g. Katz and Goodglass, 1990) and profound verbal short-term memory deficits (e.g. Martin and Saffran, 1992). The fact that the repetition errors seen in deep dysphasia and a limited number of non-fluent progressive aphasic patients are so strikingly similar may suggest common underlying functional impairments across such patients despite differences in the underlying aetiology.

On consideration of the small number of detailed studies of non-fluent progressive aphasia, it would appear that this syndrome does not necessarily comprise a homogeneous group. In particular, there is evidence to suggest differences between patients in their degree of impairment on reading, repetition and naming. This may be due to subtle differences in terms of the location of the focal atrophy in each patient. However, at present few conclusions can be drawn given that there are so few detailed reports of such patients in the literature. In this paper we present a patient, PW, diagnosed with non-fluent progressive aphasia. When repeating single words, PW made semantic paraphasias and was poorer at repeating low imageability relative to high imageability words. PW was also impaired on measures of verbal short-term memory. As outlined above, this pattern of impairment is very similar to that seen in deep dysphasia, but is rarely reported in the context of non-fluent progressive aphasia. Presently, we are only aware of two reported cases of non-fluent progressive aphasia accompanied by deep dysphasia. Given the somewhat rare nature of PW's impairment, this study will provide an additional detailed case study of language impairment in non-fluent progressive aphasia. PW's pattern of impairment will also provide an additional means of evaluating models of both primary progressive aphasia and deep dysphasia.

Case description

PW is a 60-year-old, right-handed female who left full-time education at 16 years. She has spent most of her adult life as a farmer's wife, being actively involved in the care of livestock and in day-to-day domestic activities. PW initially presented (March 1999) complaining of word finding problems, which severely impeded her ability to communicate with others, and at presentation such problems had been apparent for approximately 12 months (April 1998). Nevertheless, despite these difficulties, she continued her normal domestic day-to-day activities (i.e. cooking, cleaning, etc.) and remained a keen gardener (Bristol Activities of Daily Living 5/60; note, scoring high on this test suggests that there are problems in conducting many normal day-to-day activities). PW had been assisting in teaching at a primary school until March 1998. However, the onset of her speech production problems led to her having to resign her position.

Two initial clinical testing sessions (22 February 2000 and 15 March 2000) were conducted during which PW's performance at a variety of verbal and non-verbal tasks was examined. These testing sessions determined that PW was severely impaired on confrontational naming, reading and repetition (see Table 1). Administration of the Test for the Reception of Grammar (TROG; Bishop, 1982) revealed that grammatical and syntactic processing was also impaired.

Table 1. Basic neuropsychological data

		Norms
WAIS-III		
Verbal IQ	62	
Performance IQ	94	
BORB		
Object decision test	117/128	(115)
Item match test	31/32	(30)
Foreshortened match test	24/25	(22)
Association match test	30/30	(27)
WMS-III		
Visual reproduction II copy	100/104	(79)
Digit span (forward)	2	(6)
Digit span (backward)	2	(5)
Face recognition (immediate)	36/48	(36)
Face recognition (delayed)	34/48	(35)
Information and orientation	14/14	N/A
WRMT		
Faces	50/50	(42)
Words	48/50	(43)
Sentence comprehension		
Token Test (auditory presentation)	24/36	(33)
Token Test (written presentation)	26/36	N/A
Word–picture matching		
PALPA (auditory presentation)	40/40	(39)
PALPA (visual presentation)	40/40	(39)
Fluency		
Initial letter (FAS)	8	(36)
Animals	16	(17)
Grammatical/syntactic processing		
TROG	55/80	(78)
Picture naming		
PALPA	29/40	(39)
BNT	14/60	(56)
Word reading		
NART	5/50	(18)
Repetition		
PALPA	42/80	(79)

BNT, Boston Naming Test; BORB, Birmingham Object Recognition Battery; NART, National Adult Reading Test; PALPA, Psycholinguistic Assessments of Language Processing in Aphasia; TROG, Test for the Reception of Grammar; WAIS-III, Wechsler Adult Intelligence Scale Third Edition; WMS-III, Wechsler Memory Scale Third Edition; WRMT, Warrington's Recognition Memory Test.

PW was severely impaired at initial letter fluency. However, generation of animal names within 1 min was within the normal range. It is noteworthy that poorer performance on letter fluency relative to category fluency is a typical reported pattern in non-fluent progressive aphasia (Snowden *et al.*, 1996). Her digit span performance was consistent with a severe auditory–verbal short-term memory deficit. PW's spontaneous speech was non-fluent and agrammatic with frequent phonemic paraphasias.

Despite PW's poor performance on tests of spoken language production, word–picture matching performance was normal, although there was evidence of some sentence comprehension difficulties on the basis of both an auditory and a written version of the Token Test [De Renzi and Faglioni, 1978; two standard deviations (SD) below controls = 29]. Finally, PW performed well on object, face and word recognition tests and there was no evidence that PW had any visuospatial or perceptual deficits; she was also well orientated in space and time.

Computed tomography, magnetic resonance imaging and EEG were conducted and no abnormalities were found (all tests conducted in March 1999). However, no further examinations have since been conducted. Neurological examination of PW was unremarkable and there was no evidence of limb or speech apraxia. There was also no evidence of hearing loss and PW wears glasses for reading only. Overall, on the basis of all the clinical evidence, PW was diagnosed with non-fluent progressive aphasia.

Experimental investigations

After the initial clinical assessment, PW's language deficits were examined in greater detail. PW was visited at her home and tested in six 3-h sessions over a 9-month period (February to October 2000). Specific details concerning testing dates are provided when analysis of performance across tasks is undertaken. We have included adult norms for all tests whenever possible.

Language processing

Comprehension. The initial examination of PW's comprehension abilities involved using a word–picture matching test [Psycholinguistic Assessments of Language Processing in Aphasia (PALPA); Kay *et al.*, 1992] in which the patient is asked to point to one of five alternative pictures (which include the target and four distracters: a close semantic foil, a distant semantic foil, a visual foil and an unrelated foil that is semantically related to the visual foil). PW performed perfectly at this test when responding to both spoken and written words. Word–picture matching performance was examined further in a subsequent test (Hirsh and Funnell, 1995) consisting of 144 items in which the patient is presented with a picture and must point to the appropriate written word from an array of three words (which include the target and two distracters: a semantic foil and a phonological foil). PW performed perfectly at this task.

However, during initial clinical testing PW scored outside the normal range on both auditory and written sentence comprehension. In order to examine further PW's auditory comprehension ability, the synonym judgement task taken from the Action for Dysphasic Adults (ADA) battery was administered (Franklin *et al.*, 1992). In the first test administered all items were presented in a written form and in a second testing session all items were presented in an auditory form. As can be seen from Table 2, PW was somewhat impaired in synonym judgement regardless of test modality, and this suggests a degree of central semantic impairment. However, PW's performance was significantly worse when the items were presented in the auditory modality (McNemar change test: $\chi^2 = 4.08$, d.f. $= 1$, $P < 0.05$). In addition, on both tests, significantly more errors were made on low imageability relative to high imageability words (auditory: five high and 13 low imageability word errors; Fisher's exact test: $\chi^2 = 4.59$, d.f. $= 1$, $P < 0.05$; visual: one high and

Table 2. Test data relating to language processing

		Norms
Word–picture matching		
22 February 2000 PALPA (auditory presentation)	40/40	(39)
15 March 2000 PALPA (visual presentation)	40/40	(39)
5 April 2000 Hirsh and Funnell (1995)	144/144	(143)
Synonym judgement		
22 March 2000 ADA battery (written) high versus low imageabilty	70/80	N/A
5 April 2000 ADA battery (auditory) high versus low imageability	62/80	N/A
Lexical decision		
26 April 2000 Visual PALPA regularity	58/60	(59)
14 May 2000 Auditory PALPA regularity	56/60	(58)
26 April 2000 Visual PALPA frequency versus imageability	153/160	(158)
14 May 2000 Auditory PALPA frequency versus imageability	143/160	(158)
Reading		
22 March 2000 Howard and Franklin (1988) regularity versus imageability	195/240	N/A
22 March 2000 PALPA frequency versus imageability	42/80	(79)
5 April 2000 PALPA syllable length	60/80	(79)
26 April 2000 PALPA letter length	72/80	(79)
26 April 2000 PALPA grammatical class	38/60	(58)
26 April 2000 PALPA non-word reading	24/40 correct	N/A
Picture naming (198-item test; Morrison *et al*., 1997)		
Session one (22 March 2000)	123/198	(196)
Session two (14 May 2000)	130/198	
Session three (17 August 2000)	104/198	
Repetition		
5 April 2000 Howard and Franklin (1988) regularity versus imageability	118/240	N/A
26 April 2000 PALPA frequency versus imageability	42/80	(79)
26 April 2000 PALPA syllable length	11/24	(58)
5 April 2000 PALPA grammatical class	6/60	(39)
5 April 2000 PALPA grammatical class	9/40	N/A
5 April 2000 PALPA non-words	0/40	N/A

ADA, Action for Dysphasic Adults battery; PALPA, Psycholinguistic Assessments of Language Processing in Aphasia.

nine low imageability word errors; Fisher's exact test: $\chi^2 = 7.31$, d.f. = 1, $P < 0.01$).

Lexical decision. Given preliminary evidence that PW's reading and repetition performance was impaired, visual and auditory lexical processing were examined in two lexical decision tests taken from the PALPA test battery (Kay *et al.*, 1992). The lexical decision test involves the patient deciding whether presented items are words or non-words. The first lexical decision test administered manipulated the regularity of the real word items. As can be seen from Table 2, PW demonstrated good performance on a visual and auditory version of this test. All errors consisted of false negatives, with PW rejecting irregular words. In the second lexical decision test, the real word items were varied in terms of imageability and frequency. On the visual version of this test, PW made only eight errors, seven of which were false negatives. More errors were made on low imageability and low frequency words (two high and five low imageability errors; one high and six low frequency errors). However, these differences were not significant. On the auditory version of this test, PW performed poorly, making 17 errors (see Table 2). Fourteen of these errors were false positives, suggesting that PW is impaired in non-word processing. Overall, PW performed well on visual lexical decision but there was some marginal evidence of a poorer performance with low frequency and low imageability words. PW's performance at auditory lexical decision was poor relative to visual lexical decision, primarily due to poor non-word processing. These data suggest that PW has little impairment in processing lexical input across either modality and thus her poor performance at reading and repetition is unlikely to reflect solely a deficit in orthographic and auditory input processing. Both reading and repetition performance were examined further, and are discussed presently.

Reading. PW's reading performance was examined in several reading tests and the results of these tests are presented in Table 2. The first reading test used (Howard and Franklin, 1988) examined PW's ability to read regular/irregular words and high/low imageability words. PW made a significantly greater number of errors on irregular words relative to regular words (10 regular versus 35 irregular word errors: $\chi^2 = 7.53$, d.f. = 1, $P < 0.01$). Twelve of the errors made on irregular words were regularization errors (e.g. reading 'pint' so that it rhymes with 'lint'). She also made more errors on low imageability words (14 high versus 31 low imageability word errors: $\chi^2 = 7.90$, d.f. = 1, $P < 0.005$). The rest of the reading tests utilized were taken from the PALPA. In the first test taken from this battery we additionally examined her performance on reading high/low imageability words as well as her ability to read high/low frequency words. Although there was a trend towards an effect of frequency (15 high versus 23 low frequency word errors), it was not significant

(χ^2 = 3.21, d.f. = 1, P = 0.07). However, significantly more errors were made on low imageability words (13 high versus 25 low imageability word errors: χ^2 = 7.22, d.f. = 1, $P < 0.01$). On a test examining length in syllables, PW made significantly more errors on disyllabic words (15 two-syllable versus five one-syllable word errors: Fisher's exact test χ^2 = 6.67, d.f. = 1, $P < 0.01$). On a further test, examining length in letters, PW made significantly more errors on longer words (seven six-letter versus one three-letter word errors: Fisher's exact test χ^2 = 5.0, d.f. = 1, $P < 0.05$). PW's reading of words in different grammatical classes was examined and her performance was equally poor when reading nouns, verbs, adjectives and function words. Finally, PW's ability to read non-words was examined, and was clearly impaired (see Table 2).

Overall, PW's reading performance is somewhat similar to patients with 'surface dyslexia' (e.g. McCarthy and Warrington, 1984) in that she had greater difficulty with irregular words relative to regular words. However, unlike such patients, her non-word reading was impaired. PW's reading performance was also affected by imageability and length.

Picture naming. It was apparent that PW was impaired in picture naming on the basis of the initial clinical investigations and this was examined further in a picture naming test which consisted of 198 individual items (see Table 2). PW demonstrated poor picture naming performance in all of the sessions with this test that were undertaken. The majority of her errors consisted of circumlocutions and no responses. However, phonological and semantic errors were also present. We will examine PW's picture naming error performance in greater detail later.

In order to determine what variables affected PW's picture naming performance, we examined her ability to name a set of 198 items taken from Morrison *et al.* (1997). For all of these items we had norms on several experimental variables, and these were, imageability (scale 1–7: 1 = low, 7 = high imageability; mean = 6.2, SD = 0.38), frequency (scale 1–5: 1 = low, 5 = high frequency; mean = 2.9, SD = 0.85), age of acquisition (scale 1–7: 1 = early, 7 = late acquired; mean = 2.4, SD = 0.72), length in phonemes (mean = 4.1, SD = 1.5), visual complexity (scale 1–5: 1 = low, 5 = high complexity; mean = 2.8, SD = 0.82) and name agreement (scale 1–5: 1 = low, 5 = high agreement; mean = 4.6, SD = 0.27). For further details concerning the collection of norms on these items please refer to Morrison *et al.* (1997). Three testing sessions were undertaken over a 5-month period. A logistic regression analysis was then conducted on her naming performance at each session using the above named variables as predictors. On session one (March 2000) a significant overall R^2 was found (R^2 = 0.318, χ^2 = 75.82, d.f. = 6,192, $P < 0.0001$). The only predictors that added unique variance were frequency (change in R^2 = 0.064, χ^2 = 17.70, d.f. = 1,192, $P < 0.0001$) and length in phonemes (change in R^2 = 0.041, χ^2 = 11.66, d.f. = 1,192, $P < 0.0001$). On session two (May 2000) a significant R^2 was found (R^2 = 0.318, χ^2 = 75.8, d.f. = 6,192, $P < 0.0001$). In this case the predictors that added unique variance were age of acquisition (change in R^2 = 0.02, χ^2 = 5.85, d.f. = 1,192, $P < 0.05$), frequency (change in R^2 = 0.025, χ^2 = 7.22, d.f. = 1,192, $P < 0.01$) and length in phonemes (change in R^2 = 0.056, χ^2 = 15.64, d.f. = 1,192, $P < 0.0005$). On session three (August 2000) a significant R^2 was found (R^2 = 0.306, χ^2 = 72.39, d.f. = 6,192, $P < 0.0001$), and imageability (change in R^2 = 0.063, χ^2 = 16.78, d.f. = 1,192, $P < 0.0005$), age of acquisition (change in R^2 = 0.023, χ^2 = 6.53, d.f. = 1,192, $P < 0.01$), frequency (change in R^2 = 0.021, χ^2 = 5.97, d.f. = 1,192, $P < 0.01$) and length in phonemes (change in R^2 = 0.025, χ^2 = 6.79, d.f. = 1,192, $P < 0.01$) were all predictors that added unique variance. It should also be noted that PW made the most errors in the final testing session. An examination of error rates across testing sessions determined that performance was significantly poorer in session three relative to both sessions one and two (McNemar change test: session one versus session three χ^2 = 9.5, d.f. = 1, $P < 0.001$; session two versus session three χ^2 = 17.45, d.f. = 1, $P < 0.001$) and there was no significant difference between performance in sessions one and two. This is consistent with the degenerative nature of her illness. Overall, both frequency and length in phonemes were consistent predictors of PW's ability to name pictures.

Repetition. PW's repetition performance was examined in several tests taken from the PALPA battery. The results of these tests are summarized in Table 2. The majority of PW's repetition errors consisted of no responses. However, phonological and semantic paraphasias were also apparent, and as with her naming errors, we will discuss her error performance later. As was the case when we examined PW's reading performance, an additional test [devised by Howard and Franklin (1988)] manipulating regularity and imageability was also used. PW's performance was very poor on this test and substantially worse than her performance reading the same set of items (repetition: 118/240; reading: 195/240). Consistent with PW's reading performance, she was significantly worse at repeating low imageability words relative to high imageability words (44 high versus 78 low imageability word errors: χ^2 = 19.27, d.f. = 1, $P < 0.0001$). There was no effect of regularity on PW's repetition performance. Overall, 10% of all errors made on this test were semantic paraphasias, and they consisted of the following: body–'man'; bosom–'bust'; dove–'the pigeons'; eagle–'bird'; harem–'Arab'; malice–'nasty'; mink–'beaver'; Rabbi–'Jew'; seat–'chair'; swamp–'water'; vine–'grapes'. A second repetition test (PALPA) manipulating imageability and frequency was administered. As before, PW was significantly worse at naming low imageability words (six high versus 32 low imageability word errors: χ^2 = 33.88, d.f. = 1, $P < 0.0001$), but there was no effect of frequency (18 high versus 20 low frequency word errors). Four semantic paraphasias were also present in this test (alcohol–'beer'; lesson–'teacher'; tobacco–'cigarette'; woman–'people').

Given that PW demonstrated poor performance on disyllabic words in reading, this variable was examined in a repetition test taken from the PALPA. As it turned out, PW was equally poor at repeating polysyllabic and monosyllabic words (five three-syllable, four two-syllable and four one-syllable word errors). Two additional tests taken from the PALPA examining repetition across grammatical classes were also administered. The first test showed that PW was equivalently poor at repeating nouns and function words (16 noun and 15 function word errors). The second test showed that PW was also equally poor at repeating adjectives, verbs, nouns and function words. A final test examining non-word repetition, revealed a total inability to perform such a task. Overall, PW made semantic errors in repetition and demonstrated poor performance on repeating low imageability words and a lexicality effect.

Error performance across language tasks. Our testing of PW's performance on tests of reading, repetition and picture naming determined that she was impaired on all such tasks. Examination of PW's performance across these tasks (see Table 2) suggests that her impairment on repetition tasks was possibly greater relative to her impairment at reading and picture naming. However, this conclusion is based on a performance that in many cases involved different sets of items. For this reason, PW's performance on all three tasks was examined with the same set of 198 items (Morrison *et al.*, 1997) that had been employed to examine her picture naming performance. In this case, PW's ability to read and repeat these items was examined shortly after naming session three (naming: 02 August 2000; reading: 16 August 2000; repetition: 30 August 2000). Error performance across all the naming tests administered with these items was also examined and the results are summarized in Table 3. Finally, as an extra comparison of repetition and reading performance, error rates on the respective versions of the Howard and Franklin (1988) test are also included in Table 3. Errors were classified into nine major types and these classifications are based on those proposed by Martin and Saffran (1992). These categories have also been employed in other studies of non-fluent progressive aphasia (Croot *et al.*, 1998; Majerus *et al.*, 2001). The categories were: (1) formal paraphasias which are responses that are phonologically but not semantically related to the target response (e.g. thimble–'nimble'), (2) semantically related errors which are subdivided into (a) semantic paraphasias which are responses consisting of a single semantically related word (e.g. glove–'hand') and (b) semantic descriptions or circumlocutions that are semantically related to the target response (e.g. anchor–'in the boat and go in the water'), (3) formal and semantically related paraphasias or mixed errors (e.g. ladder–'stir' via 'stairs'), (4) unrelated lexical errors which are lexical responses that bear neither a semantic nor a phonological relation to the target (e.g. trumpet–'boy'), (5) target-related neologisms which are non-

Table 3. Error response types across all tasks (% responses given)

	198-item set						Howard and Franklin (1988) set		
Response type	Naming session one March 2000	Naming session two May 2000	Naming session three August 2000	Repeat words August 2000	Read words August 2000	Total	Repeat words March 2000	Read words April 2000	Overall total
Target correct	62	65	52	47	64	58	49	81	60
Formal paraphasia	1	2	4	6	10	4	9	6	5
Semantic paraphasia	2	2	1	6	0	2	5	0	2
Semantic description	10	7	7	3	0	6	0	0	4
Formal and semantic paraphasia	0	0	1	3	1	1	0	1	1
Neologism on a semantic paraphasia	0	0	1	2	0	1	0	0	1
Target-related neologism	6	8	6	10	23	10	6	11	10
Abstruse neologism	0	0	1	1	2	1	1	0	1
Unrelated lexical	2	0	4	1	0	2	2	0	1
No response	17	15	22	21	0	15	28	1	15

words that sound similar to the target-related response (e.g. dummy–'denny'), (6) neologism on a semantic paraphasia which is a response that is a non-word that sounds similar to a semantic neighbour of the target response (e.g. glasses–'SpEktl' via 'spectacles'), (7) abstruse neologism which is a response that is a non-word that is totally unrelated to the target response (e.g. equal–'wuk'), (8) no response errors. In line with Martin *et al.* (1994), our criteria for the classification of a formal paraphasia error were fairly liberal: the error had to be a word and had to have at least one consonant or stressed vowel in common with the target. Target-related neologisms were selected in the same manner, but in this case all responses had to be non-words. Semantically related errors were judged to be the case by three undergraduate students who were presented with the entire corpus of PW's errors and asked to select which (if any) of the items were related in terms of meaning.

If we examine PW's correct response rate on the same set of 198 items across production tasks (see Table 3), it is clear that both repetition (47% correct) and naming (52% correct) were poor relative to reading (64%). Overall, reading performance was significantly better than both repetition (McNemar change test: $\chi^2 = 15.83$, d.f. $= 1$, $P < 0.001$) and naming (McNemar change test: $\chi^2 = 8.11$, d.f. $= 1$, $P < 0.001$). There was no significant difference between repetition and naming performance. Consistent with the earlier findings, repetition was significantly poorer than reading on the Howard and Franklin (1988) test (McNemar change test: $\chi^2 = 60$, d.f. $= 1$, $P < 0.0001$). The pattern of errors also differed across production tasks. On reading tasks, target-related neologisms were the most common error type, followed by formal paraphasias. On naming tasks, no responses were most common, followed by semantic description errors and target-related neologisms. On repetition tasks, no response errors were once again the most common error type, but several other types of error were also present, including target-related neologisms, semantic paraphasias and formal paraphasias.

Summary. The results of the language-based testing indicated the following: (a) PW performed well on measures of comprehension, but showed significantly poorer performance when the items were presented orally; (b) PW demonstrated little impairment on visual lexical decision but there was early indication of poorer performance on low imageability words. It is also noteworthy that lexical decision performance was superior to reading on the same list of words (PALPA frequency versus imageability: visual lexical decision 73/80; reading 42/80); (c) PW demonstrated some poor performance on auditory lexical decision with particularly poor performance with non-word items and no evidence of imageability effects. Nevertheless, auditory decision performance was superior to repetition on the same list of words (PALPA frequency versus imageability: auditory lexical decision 79/80; repetition 42/80); (d) PW's reading performance was poorer with exception words, and she was also significantly worse with low imageability words and disyllabic words; (e) PW's poor picture naming performance was consistently influenced by length in phonemes and word frequency; (f) PW's repetition performance was significantly worse with low imageability words and totally abolished for non-words; (g) a comparison of performance across production tasks determined that both repetition and naming performance were significantly more impaired than reading. PW also made semantic errors in repetition and naming but no such errors in reading. Overall, on the basis of repetition and reading performance, it was apparent that PW's processing of verbal input was poorer when information was presented in the auditory modality relative to the visual modality. Given that auditory lexical decision performance was relatively spared, it is unlikely that this difference in performance is exclusively due to an impairment in auditory input processing, but this possibility was examined further. Alternatively, this difference in performance could reflect an impairment in auditory–verbal short-term memory. In order to explore these two possibilities, two additional sets of tests were conducted. The first set of tests examined PW's auditory phonological input processing and general phonological awareness. The second set of tests examined PW's auditory–verbal short-term memory.

Tests of auditory phonological processing

Phoneme discrimination. In order to determine whether PW's poor performance at repetition was due to an impairment in auditory phonological input processing, a series of phonological discrimination tests was administered. Two auditory discrimination tests were taken from the PALPA battery. The first test (known as a minimal pairs test) involved making same/different judgements on pairs of monosyllabic consonant-vowel-consonant words, which were identical or differed by a single feature of place or voicing (e.g. pact–fact). The second test was identical except that non-word items were presented (e.g. beck–feck). PW was well within the normal range on both tests (see Table 4). A second set of discrimination tests (N. Martin, personal communication) was conducted to explore the effects of delay on same/different judgements of pairs of words and pairs of non-words. In the first test, PW was presented with word pairs and non-word pairs with no delay between the presentation of the first and the second item of the pair. The second test involved a 5-s interval between presentations. The final test also involved a 5-s interval between item presentations, but PW was additionally asked to count from one to five during this interval. In all cases the tests were administered in such a way that lip reading was not possible. PW performed within the normal range on all three tests (see Table 4).

A final test of phoneme segmentation (Muter *et al.*, 1997) was also conducted in which the patient must take the initial sound away from a presented word and say what remains (e.g. say 'cow' without the first sound). PW performed well on this task scoring 13/14 correct with one no response error.

Table 4. Test data relating to phonological processing

		Norms
Phonemic discrimination		
29 June 2000 PALPA same/different judgement of word pairs	69/72	(70)
29 June 2000 PALPA same/different judgement of non-word pairs	68/72	(70)
27 September 2000 minimal pairs words/non-words		
Test 1 (no delay)	32/32	(32)
Test 2 (unfilled delay)	31/32	(32)
Test 3 (filled delay)	30/32	(32)
Rhyme judgement (auditory presentation)		
5 April 2000 PALPA test 1	33/40	N/A
26 April 2000 PALPA test 2	55/60	N/A
Rhyme judgement (written presentation)		
14 May 2000 PALPA test 1	20/40	N/A
24 May 2000 PALPA test 2	37/60	N/A
24 May 2000 PALPA non-words	34/50	N/A
Homophone judgement (written presentation)		
24 May 2000 PALPA regular homophones	38/50	N/A
24 May 2000 PALPA irregular homophones	38/50	N/A

PALPA, Psycholinguistic Assessments of Language Processing in Aphasia.

Overall, PW performed normally on phoneme discrimination and segmentation tests, which suggests that she has no auditory input processing impairment.

Rhyme judgement. A series of rhyme judgement tests (taken from the PALPA battery) was administered to PW in order to determine whether there was any general impairment in phonological processing. It has been argued that in order to carry out a rhyme judgement task, both items must be held in storage temporarily while the relevant parts of the words are segmented and compared (Monsell, 1987). Thus, these tests should help us to determine whether PW's poor repetition performance should be attributed to a deficit in short-term storage of phonological information. Both rhyme judgement tests involved auditory presentation of rhyming and non-rhyming pairs of items with PW indicating whether the items did or did not rhyme, and crossed with rhyme status was visual similarity. Thus, both tests involved selecting rhyme and non-rhyme items from four conditions, which included visually similar rhymes (e.g. town–gown), visually dissimilar rhymes (e.g. ghost–roast), visually similar non-rhymes (e.g. cheat–sweat) and visually dissimilar non-rhymes (e.g. flair–year). Although the items used in each list were different they did not differ in difficulty. PW demonstrated a less than perfect performance at both tests, making both false-positive and false-negative errors (test 1: three false-negative and four false-positive errors; test 2: five false-negative errors). There was no effect of visual similarity. Thus, these results suggest that PW has an impairment of short-term phonological storage.

Given that PW's reading performance suggested an impairment in access to appropriate phonology from orthographic input, a series of visual rhyme judgement tests was administered. The first two rhyme judgement tests were visual versions of the PALPA tests described above. PW performed poorly on both tests and the majority of errors on both tests were false positives (test 1: seven false-negative and 13 false-positive errors; test 2: seven false-negative and 16 false-positive errors). A third rhyme judgement test was also administered in which all items were non-words, and PW similarly performed poorly (see Table 4). Interestingly, the majority of the false-positive errors PW made on the first two tests were to visually similar non-rhyme items in which she inappropriately selected (test 1: 10/13 false-positive errors; test 2: 13/16 false-positive errors). Overall, PW performed significantly worse at visual word rhyme judgement relative to auditory word rhyme judgement (McNemar change test: test 1: $\chi^2 = 8.64$, d.f. = 1, $P < 0.001$; test 2: $\chi^2 = 12.04$, d.f. = 1, $P < 0.001$). In particular, PW was almost entirely unable to reject visually similar non-rhyme items. Such a pattern of performance has been reported previously in a patient with non-fluent progressive aphasia (case NL; Dowhaniuk *et al.*, 1999) who despite unimpaired reading and comprehension, made a great number of false-positive errors with visually similar rhymes.

Homophone judgement. Given PW's poor performance on visual word rhyme judgement, an additional set of homophone judgement tests was administered. Homophone judgement involves the visual presentation of orthographically dissimilar pairs of items that may or may not sound the same (e.g. raise–rays; home–him). Homophone judgement differs from rhyme judgement in that no phonological segmentation process is needed. In this case the whole word phonological representations (or lexemes) corresponding to each item in the pair are retrieved, stored and compared without the additional need to segment particular relevant parts. Thus, this task would demonstrate if PW had deficits in access and short-term storage of appropriate phonology from orthographic input above and beyond possible impairments in phonological segmentation. In this case all the items in both homophone judgement tests were orthographically dissimilar, and thus PW could not rely on visual similarity in performing this task. All homophones in the first test were regular and in the second test all homophones were irregular. Both tests are taken from the PALPA testing battery. As can be seen from Table 4, PW performed very poorly on both tests, and as was the case with rhyme judgement, the majority of her errors were false positives (test 1: 12 false-positive errors; test 2: 10 false-positive and two false-negative errors). PW was also equally poor at regular and irregular homophone judgements. Overall, the results of both tests provide further evidence that PW is impaired in retrieving appropriate phonology from orthographic input and furthermore that her short-term storage of such information is also potentially impaired. This possibility was explored in the next set of experiments.

Summary. PW was impaired on written rhyme judgement and homophone judgement tasks, making a large number of false-positive errors in both cases. On the written rhyme judgement tasks, the majority of these false-positive errors were to visually similar non-rhyme items (e.g. pint–lint). This result is consistent with her poor irregular word reading performance, and further demonstrates her inability to retrieve appropriate phonology from orthographic input.

PW performed normally on tests of auditory discrimination and segmentation, which suggests that she has no impairment in auditory input processing. However, PW was mildly impaired in auditory rhyme judgement, although written rhyme judgement was significantly worse. Nevertheless, this may not reflect any underlying impairment in auditory phonological processing *per se*, and could be attributed to an impairment in auditory–verbal short-term memory. An auditory–verbal short-term memory impairment could also explain PW's poor repetition performance and this issue was explored in the next set of tests.

Short-term and long-term memory processing

Auditory–verbal short-term memory. Several measures of auditory–verbal short-term memory were administered to PW. An initial clinical examination of PW found that she was only able to repeat two items on a digit span task. On a similar task involving the repetition of sequences of letters and numbers, PW's performance was again limited to just two items. Both these results suggest that PW has a severe deficit of auditory–verbal short-term memory. However, in order to rule out the possibility that poor performance on digit span was due to an impairment in accessing phonological output representations, two matching span tasks were conducted. In the first matching span task [taken from the Wechsler Adult Intelligence Scale-III (WAIS-III)] the patient is asked to indicate whether pairs of digit sequences of increasing length are the same or different (e.g. '4, 6, 7' and '6, 4, 7'). Items are presented at a rate of one per second, with a 3-s pause between each presented sequence. The second matching span task (taken from the PALPA) used the same procedure, but words were presented instead of digits. PW performed very poorly on both tests (see Table 5) and was only able to match successfully a sequence of two items. Overall, the results of these tests suggest that PW has a severe impairment of auditory–verbal short-term memory.

Table 5. Test data relating to short-term and long-term memory processing

Auditory–verbal short-term memory	
22 February 2000 WAIS-III digit span (forward)	2
22 February 2000 WAIS-III digit span (backward)	2
22 February 2000 WAIS-III letter–number sequencing	2
15 March 2000 PALPA digit matching span	2
15 March 2000 PALPA word matching span	2
Long-term memory	
Hopkins Verbal Learning Test	
Session one (22 March 2000) Auditory presentation and spoken responses	
Free recall: trial 1: 0/12; trial 2: 3/12; trial 3: 6/12	
Auditory recognition: true positives: 6/12; false positives: 0/12	
Session two (5 April 2000) Auditory presentation and written responses	
Free recall: trial 1: 2/12; trial 2: 3/12; trial 3: 5/12	
Auditory recognition: true positives: 5/12; false positives: 0/12	
Session three (26 April 2000) Visual presentation and spoken responses	
Free recall: trial 1: 8/12; trial 2: 9/12; trial 3: 11/12	
Visual recognition: true positives: 12/12; False positives: 0/12	
Session four (14 May 2000) Visual presentation and written responses	
Free recall: trial 1: 6/12; trial 2: 10/12; trial 3: 11/12	
Visual recognition: true positives: 12/12; false positives: 0/12	

WAIS, Wechsler Adult Intelligence Scale Third Edition; PALPA, Psycholinguistic Assessments of Language Processing in Aphasia.

Long-term memory. PW showed consistently poorer performance on auditory relative to visually presented language tasks (see Table 2). This pattern of performance probably reflects a central deficit in general auditory–verbal short-term memory. In order to determine the impact of this deficit on the encoding and storing of verbal material in long-term memory, PW's performance at free recall of lists of words across modalities was examined. The test utilized was the Hopkins Verbal Learning Test (Brandt, 1991) which involved presenting PW with a list of 12 items from three semantic categories (e.g. four-legged animals, precious stones and human dwellings) with four items corresponding to each category. After each presentation trial PW was asked to free recall as many items from the list as she could. This test was run on four separate occasions (with 2–3 weeks between each testing occasion) in order to manipulate the modality in which items were presented (i.e. auditory or visual presentation) and the modality in which responses were to be made (i.e. spoken or written responses). A different list of 12 items was used for each testing session in order to prevent the possibility of confounding effects of learning across sessions. All four lists were taken from the Hopkins Verbal Learning Test and all lists have been demonstrated to be equivalent in difficulty (Brandt, 1991). When items were presented in the auditory modality, the experimenter read aloud each word on the list at a rate of one every 2 s. When items were presented in the visual modality, the experimenter presented each word on a card (written in Times New Roman font; 48 point). The results of these testing sessions are summarized in Table 5. PW performed better when the items were presented in the visual modality relative to the auditory modality, regardless of whether spoken or written responses were required. After each recall test an additional recognition test was conducted at each of the four testing sessions. The recognition test consisted of 24 items, half of which had been presented in the recall test.

The 12 distracter items consisted of six items which were semantically related to items on the recall list and six unrelated items (Brandt, 1991). The recognition items were presented either visually or orally and consistent with her responses in the recall test, PW responded with written responses (i.e. a tick or cross) or verbally (i.e. 'yes' and 'no'). From Table 5 it may be seen that the visual recognition performance was consistently better than the auditory recognition performance, regardless of response modality. Overall, these results are entirely consistent with the proposal that PW's storage and subsequent retrieval of auditory phonological input is substantially impaired.

Summary. PW demonstrated a very severe impairment in auditory–verbal short-term memory. This deficit was apparent in both span and matching tasks. There was also evidence of poorer recall and recognition performance when items were orally presented relative to visually presented. Overall, given that there was no evidence of an impairment in auditory phonological processing on the basis of tests of auditory discrimination and segmentation, it is likely that a severe impairment of auditory–verbal short-term memory underlies the differences seen in PW's performance across modalities in comprehension testing and may also explain PW's poor repetition performance. Interestingly, a severe deficit of auditory–verbal short-term memory has also been used to explain the pattern of repetition performance seen in patients diagnosed with deep dysphasia. Given that PW's impairment in repetition is so similar to that seen in deep dysphasia, this aspect of her performance will be considered in the context of current models of that disorder.

General discussion

In this study we examined the language performance of a single patient, PW, diagnosed with non-fluent progressive aphasia. Consistent with this diagnosis, PW's spontaneous speech was non-fluent and agrammatic. However, our testing showed that PW was unimpaired on several measures of non-verbal performance. PW was also relatively unimpaired on comprehension testing, but did show significantly poorer performance when items were presented orally. She performed poorly on tests of reading, repetition and picture naming. There was also evidence that picture naming performance was deteriorating over time, with significantly poorer performance on her last testing session relative to earlier testing sessions. A closer examination of performance on each of the different production tasks indicated that: (1) PW's performance in picture naming was influenced by length in phonemes and word frequency; (2) PW's reading performance was significantly poorer with exception words, low imageability words, disyllabic words and non-word reading was impaired; (3) PW's repetition performance was significantly poorer with low imageability words and she was entirely unable to repeat non-words; (4) word reading was significantly less impaired than both repetition and naming, whereas there was no significant difference in repetition and naming performance.

Given the evidence that PW's performance was poorer on orally presented words relative to visually presented words, both auditory phonological processing and auditory–verbal short-term memory were examined. There was no evidence of an impairment in auditory phonological processing as PW performed normally on tests of auditory discrimination and segmentation. However, PW was very impaired on measures of auditory–verbal short-term memory, with a very poor performance on both digit matching and span tasks. Overall, several aspects of PW's pattern of performance are strikingly similar to the aphasic syndrome known as 'deep dysphasia'. Consistent with the four major features of such patients (outlined earlier): (1) PW made semantic paraphasias in repetition; (2) PW was totally unable to repeat non-words; (3) PW demonstrated imageability effects in repetition; (4) PW performed poorly at measures of auditory–verbal short-term memory. Therefore, it has been clearly established that this aspect of her aphasic profile is consistent with deep dysphasia. As mentioned previously, there have only been two other reports of deep dysphasic performance in the context of non-fluent progressive aphasia (Croot *et al.*, 1999; Majerus *et al.*, 2001). In both studies, the authors explain their patient's performance in the context of the model of deep dysphasia proposed by Martin and Saffran (1992).

Martin and Saffran (1992) proposed an interactive model of speech production to account for the performance of their deep dysphasic patient NC [see also Martin *et al.* (1994, 1996)]. Their model includes three layers of representational units consisting of phonological, lexical and semantic representations [this model is adapted from an earlier model of normal speech production first proposed by Dell (1986)]. The model postulates that damage can occur in two major ways, either weakening of connection strength (i.e. the links between units across representational layers via which activation is passed) or an abnormal increase in decay rate (i.e. the speed with which activation of any given unit returns to its resting level). In a task like repetition, it is assumed that phonological units increase in activation first (as the input is auditory in nature) and these subsequently feed activation forward to lexical and semantic units. For example, if the input is the word 'dog' this will increase the phonological units corresponding to this word (i.e. /d/, /o/ and /g/) and in turn the relevant lexical and semantic units corresponding to this word will increase. However, at the same time, other words that share several of these phonological units (e.g. 'log', 'bog', 'dot', etc.) will also become active, resulting in an increase in the activation levels of the semantic and lexical units corresponding to these words. Subsequent feedback from semantic and lexical features will occur and further increase the initial activation of all previously active phonological units. Given the assumption that activation feeds forwards and backwards between phonological, lexical, and semantic representations, the target phonological output typically will receive the most activation and be selected. However, assuming normal connection strength, if the decay rate is uniformly increased (as a result of 'damage' to the network) this will have a significant effect on units that have been activated early by an incoming input (at any given representational level) relative to units activated later (as a result of feed-forward from these earlier activated units). In the above example, if the activation of the phonological units corresponding to the initially presented word decreases substantially, these units will no longer have any advantage when feedback occurs from the semantic units. Given that in this case, feedback from semantic units would result in the activation not just of the target's phonological representations but those also corresponding to semantically related words, this can increase the probability of selection of a semantically related item and thus a semantic error in the repetition task.

Martin and colleagues found that manipulating the decay rate of their model resulted in a pattern of performance that was identical to the repetition performance demonstrated by NC (who in repetition made a larger number of semantic errors relative to all other types of lexical error). An increase in decay rate can also explain why NC made a larger number of phonologically related neologisms and formal errors in picture naming relative to all other error types. In a task like confrontational naming, it is the semantic units that will become active first, feeding this activation forward to phonological representations. Assuming rapid decay of these initially active semantic units, this will lead to an increase in the probability of a phonologically related competing item being selected inappropriately. Finally, Martin and colleagues point out that a pathological decay rate can also explain NC's poor performance on measures of auditory–verbal short-term memory: namely, that the target's phonological representations do not remain active long enough in a short-term 'buffer' for appropriate encoding to occur, thus resulting in no responses. Overall, Martin and colleagues provide an impressive model within which all their patient's deficits in performance can be accounted for by the manipulation of a single parameter.

As mentioned previously, this model has also been used to account successfully for the deficits in performance reported in non-fluent progressive aphasic patients. Croot *et al.* (1998) concluded that performance in their two patients, PG and LM, was best simulated within the model by assuming a pathological weakening of connections between representational units. The pattern of output associated with such damage to the model consists mainly of neologistic errors and a higher probability of semantic and lexical errors in picture naming than in repetition and reading, which is consistent with their patients' performance. In a subsequent study, Croot *et al.* (1999) describe brothers diagnosed with non-fluent progressive aphasia. Although initially CB and RB showed a pattern of poorer performance at naming tasks relative to repetition and reading, the brothers did not show the same pattern of performance over time. Overall, RB showed a pattern of impairment that was similar to PG and LM, and they concluded that a 'weakened links' explanation was also suitable as an account of this patient's deficits. However, unlike his brother, CB demonstrated both imageability effects and semantic errors in repetition. As discussed previously, this pattern of performance is similar to that reported in deep dysphasia. Consistent with the Martin and Saffran (1992) account of such patients, Croot and colleagues concluded that CB's performance is best explained by an increase in decay rate. In the only other subsequent report of deep dysphasia in the context of non-fluent progressive aphasia, the authors similarly concluded that performance in their patient (CO) was also consistent with an abnormal decay rate explanation (Majerus *et al.*, 2001). A brief summary of the performance of each of these patients is provided in Table 6.

Given that we have established that PW demonstrated both imageability effects and semantic errors in repetition, her performance is consistent with two patients mentioned above (CB: Croot *et al.*, 1999; CO: Majerus *et al.*, 2001). As was the case with both of these patients, PW was also impaired attempting to repeat non-words. PW's performance across production tasks is also very similar to other previous detailed reports of non-fluent progressive aphasia. PW's performance is consistent with both patients (LM, PG) reported by Croot *et al.* (1998), in that her reading performance generated significantly more correct responses than repetition and naming. PW is also consistent with one of these patients (LM) in that she showed no significant difference in the number of correct responses across repetition and naming (see Table 6). However, PW differs from the patients (CB and RB) reported by Croot *et al.* (1999) in that both of these patients demonstrated consistently better performance at reading and repetition relative to naming. Unfortunately, direct comparison of error distributions across patients is not possible, as they were not matched for severity of deficit. Nevertheless, if we tentatively compare PW's reading and repetition performance with that of CB (the only patient from this group who made outright semantic errors in repetition), we see that their error performance on reading tasks is also similar in that the majority of errors were either target-related neologisms or formal paraphasias. However, CB and PW differ in naming error performance. The majority of PW's errors were no responses and semantic descriptions, whereas the majority of CB's errors were target-related neologisms. A comparison of PW's performance across production tasks with that of CO is more favourable (see Table 6). Consistent with PW, CO demonstrated superior reading performance

Table 6. Comparison of performance across patients

Patient	Performance across tasks	Impaired non-word repetition	Impaired non-word reading	Semantic errors in repetition	Imageability effects in repetition
PG	Reading > repetition > naming	Yes	Yes	No	N/A
LM	Reading > repetition = naming	Yes	Yes	No	N/A
RB	Repetition > reading > naming	Yes	Yes	No	No
CB	Reading > repetition > naming	Yes	Yes	Yes	Yes
CO	Reading > repetition = naming	Yes	No	Yes	Yes
PW	Reading > repetition = naming	Yes	Yes	Yes	Yes

PG, LM: Croot *et al.* (1998); RB, CB: Croot *et al.* (1999); CO: Majerus *et al.* (2001).

and was equivalently impaired on reading and repetition. An examination of CO's error performance in both reading and naming is also consistent with PW in that the majority of his reading errors were phonemic. Consistent with PW, CO's reading errors were predominately formal paraphasias and the majority of his naming errors were no responses. Thus it would appear that some of the previously reported patients (LM, PG and CO) are consistent with PW in terms of general performance across production tasks, in that all show better performance at reading relative to repetition and naming in terms of numbers of correct responses. However, superficial examination of the types of error made in each production task across the two other patients (CO and CB) who demonstrated a similar pattern of repetition performance, suggests that at least in the case of CB performance is not entirely consistent with that of our patient PW.

Given the fact that the performance of both CO and CB has been explained by assuming a pathological rate of decay activation and the fact that PW's repetition performance is similar to both of these patients, we will conclude that her repetition performance may also be best explained by the pathological decay hypothesis. However, adopting this explanation as a means of explaining her performance at naming and reading tasks is somewhat problematic. The pathological decay hypothesis makes the prediction that picture naming errors should see a gradual increase in the proportion of formal errors (e.g. thimble–'nimble') and unrelated lexical errors (e.g. trumpet–'boy') relative to semantic errors (e.g. glove–'hand') as performance worsens. This increase in formal and unrelated lexical errors is largely due to feedback from phonological representations coupled with the abnormal decay of the semantic representations corresponding to the previously active target, which leads to an increase in the probability that phonologically related and eventually unrelated words will be selected inappropriately (Martin and Saffran, 1992). In line with this prediction, there was a small increase in the percentage of formal paraphasias and unrelated lexical responses made in PW's picture naming by session three (see Table 3). However, PW made a large number of no response errors in naming as well as a large number of semantic description errors. Unfortunately, the most recent model of aphasic naming performance proposed by Dell *et al.* (1997) is limited to patients who make a total of less than 15% of both these types of error. For this reason it is currently not possible to simulate PW's performance adequately within this model, and thus impossible to determine whether the pathological decay hypothesis is indeed the most apt. Nevertheless, the pathological decay hypothesis could also potentially explain why PW's reading performance was superior to her repetition performance. If we assume that visual input (i.e. the written word) in a reading task provides a less transitory source of activation that serves as an additional advantage that is not present for auditory input (i.e. the spoken word), this could offset the effects of abnormal decay. This advantage of visual input may be enough to facilitate the appropriate naming of the target words, but not enough to ensure perfect performance. At present this is just conjecture and it still remains unclear whether this would reflect a benefit of activation that is specific to the phonological or semantic features of the target word (or both). Nevertheless, if we assume that reading a written word results in prolonged activation at the level of phonological representations (as the stimulus remains present throughout reading), this advantage would prevent semantically related items gaining higher phonological activation in the event of abnormal decay. In a task like repetition the stimulus is more transitory, and thus there is no potential for an effect of prolonged phonological activation to offset the effects of abnormal decay. Such an explanation could explain why PW (and patients like her) made semantic errors exclusively in repetition and not in reading (as seen in deep dyslexia). However, it remains unclear how the abnormal decay hypothesis can explain why PW made a greater number of errors on irregular words relative to regular words (i.e. surface dyslexia). Unfortunately, neither the models proposed by Dell and colleagues, nor those proposed by Martin and colleagues, have attempted to provide an account of reading performance. For this reason we cannot speculate as to how the pattern of PW's reading deficit can be explained. Nevertheless, we will continue to monitor the pattern of decline in PW's performance on reading relative to repetition and naming tasks in the hope of obtaining evidence which may indicate how the models proposed by Dell and colleagues and Martin and colleagues (and other interactive activation-type models like them) can be appropriately modified. For the moment we must look elsewhere to explain PW's reading deficits.

In a series of tests it was established that PW made more errors reading irregular words relative to regular words and was poor at reading both low imageability and disyllabic words. The regularity effect shown by PW is similar to that seen in 'surface dyslexia', and such effects have been reported in some patients with non-fluent progressive aphasia (Watt *et al.*, 1997; Noble *et al.*, 2000). PW also made more false-positive errors on visually similar items in rhyme judgement tasks (e.g. 'pint'–'lint'), and this is consistent with her surface dyslexic reading performance, in that such errors can occur when irregular words are inappropriately regularized. Surface dyslexia has been explained within dual-route models of reading (Funnell, 1983; McCarthy and Warrington, 1984; Marcel, 1987; Ellis and Young, 1988; Coltheart *et al.*, 1993) by assuming that the lexical reading route is damaged in some way. The outcome of an over-reliance on the sublexical reading route would be regularization errors with irregular letter strings being converted to incorrect phonemic strings on the basis of regular words' grapheme–phoneme correspondences. Nevertheless, reading via the sublexical route should result in normal reading of both regular words and non-words [for further details see Coltheart *et al.* (2001) for an excellent review of dual-route models of reading].

Although PW's reading performance is similar to surface dyslexia in terms of her poor performance with irregular words, unlike such patients she was also impaired at reading non-words. Thus, with reference to a dual-route model of reading, PW's non-word reading performance would suggest that it is unlikely that her success at reading regular words was entirely due to reading via the sublexical route. Additionally, less than half of her errors on irregular words were regularization-type errors, and this is further evidence that PW's performance was not entirely dependent on the sublexical route. Given that PW was able to do visual lexical decision, we would argue that she is able to access stored orthographic representations. PW also performed well on written word tests of comprehension, which implies normal access to semantic representations from orthography. However, her reading performance was poor and thus access to phonological output representations must be disrupted in some manner.

Recently, Coltheart *et al.* (2001) proposed a dual-route cascaded model of reading which has been computationally implemented. This model can provide an account of PW's non-word reading deficits by claiming that it reflects damage to the non-lexical grapheme–phoneme system. However, given that PW's also made errors reading real words, there must also be damage to the lexical reading route. Let us consider how partial damage to both lexical and sublexical routes could be used to provide an account of PW's reading performance within the model proposed by Coltheart and colleagues. On our non-word reading list PW scored 24/40 which suggests 60% correct via the non-lexical reading route. When reading the words from the test devised by Howard and Franklin (1988) she scored 85/120 on irregular words which suggests 71% correct via the lexical reading route. Given that the regular words list from this test is matched with the irregular words list, 71% of the time PW will be able to read the regular words lexically (therefore also scoring 85/120). However, when regular words cannot be read via the lexical route, the non-lexical route may also be employed. Given that we assume that the non-lexical route operates normally 60% of the time, we would suggest that this proportion of regular words that cannot be read lexically will be read correctly via the non-lexical route. Thus, if PW can read 85/120 regular words via the lexical route, for the 35 words she cannot read lexically, 60% (or 21 words) will be correct when read using the non-lexical route. Thus, the model predicts PW will score $85 + 21 = 106$ regular words correct. In fact this is very close to her actual score of 110/120 correct on the list of regular words[2]. Clearly, on the basis of this example it would appear that PW's reading performance can potentially fit what would be predicted by a dual-route reading model. However, we intend to conduct future work with PW to examine the pattern of decline in her reading performance in terms of degenerative damage to each of these prospective reading routes.

Given that we have utilized a dual-route model to provide an account of PW's reading performance, in the interests of consistency we shall consider such models regarding her deficits in repetition and naming. We should stress that the models we will consider at this point are in the main descriptive, and thus such models can only identify potential locations for damage rather than making testable predictions that can be examined in the light of an implemented version of the model. In this paper we have focused on the models of Dell and colleagues and Martin and colleagues as they provide a more specific account with which we have examined at least two aspects of PW's language deficits (i.e. repetition and naming). For this reason our consideration of such models as a means of explaining PW's deficits in repetition and naming will be brief.

Several non-interactive modular models have been proposed to provide an account for the processes underlying reading, repetition and naming (Patterson and Shewell, 1987; Howard and Franklin, 1988; Ellis and Young, 1988; Hillis and Caramazza, 1994). The routes available to fulfil repetition tasks mirror those we have outlined for reading tasks in that two repetition routes are proposed, one of which is sublexical (acoustic-to-phonological conversion) and the other lexical, with processing occurring from an auditory input lexicon to the phonological output lexicon via the semantic system. Given that PW was heavily impaired in non-word repetition and made semantic errors in repetition, her pattern of performance would be consistent with a dependence on the lexical–semantic route to repetition with the sublexical repetition route being severely damaged. In the case of picture naming, non-interactive modular models propose that only a single route is available to fulfil such a task, with access to phonological representations only occurring via the semantic system. Given that PW's pattern of picture naming performance is predominately anomic her deficit would be consistent with damage to the links between representations within the semantic system and their corresponding representations in the phonological output lexicon. Overall, an account of PW's deficits in repetition and naming within non-interactive modular models would involve severe damage to the sublexical repetition route coupled with damage to the links between the semantic system and the phonological output lexicon. It should also be noted that PW performed relatively well on auditory discrimination and segmentation tasks which suggests that her auditory input system is preserved (see Table 4). Whereas, PW's good performance on the tests from the Birmingham Object Recognition Battery (see Table 1) suggests that her object recognition system is also well preserved.

In conclusion, our study of PW provides further evidence as to the nature of language impairment in non-fluent progressive aphasia. Consistent with two other reported cases, our patient demonstrated a profile that is consistent with that seen in deep dysphasia. However, such a pattern of performance is rarely reported in the context of non-fluent progressive aphasia and suggests that a small subset of such patients present with a severe deficit in repetition. PW's performance was considered with reference to the connectionist model of deep dysphasia proposed by Martin and colleagues [which is consistent with the model proposed by Dell *et al.* (1997)]. It was concluded that her pattern of repetition impairment could be predicted by an underlying pathological decay in activation within this model. Unfortunately, PW's performance at both naming and reading is currently beyond the scope of this model. Overall, PW demonstrated superior reading performance relative to naming and repetition, but equivalent performance on naming and repetition and this is consistent with other patients reported within the literature (PG, LM: Croot *et al.*, 1998; CO: Majerus *et al.*, 2001). We conclude that differing patterns of repetition performance across non-fluent progressive aphasic patients are consistent with either pathological decay (PW, CO, CB), with such patients demonstrating a repetition impairment consistent with deep dysphasia, or pathological weakening (PG, LM, RB) within the speech production model proposed by Dell *et al.* (1997) and Martin *et al.* (1994). However, at present this conclusion is based only on a handful of detailed reports of non-fluent progressive aphasia, and therefore further evidence is clearly needed.

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Notes

[1]It is possible that an unsuitably high proportion of the items used in our 198 picture naming test were one-syllable words and this would have increased the likelihood that formal errors would have occurred. However, a comparison of our test with the Philadelphia Naming Test (PNT) on which Dell's model is based demonstrates that this is unlikely to be the case. In our test, 57% of the items were single-syllable words, 34% were two-syllable words, 8% were three-syllable words and 1% were four-syllable words, whereas in the PNT, 58% of items were single-syllable words, 28% were two-syllable words, 12% were three-syllable words and 2% were four-syllable words. We would like to thank an anonymous reviewer for making this point.

[2]We would like to thank Max Coltheart for his helpful comments regarding this analysis of PW's reading performance.

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Deep dysphasic performance in non-fluent progressive aphasia: a case study

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Abstract

We present a patient (PW) with non-fluent progressive aphasia, characterized by severe word finding difficulties and frequent phonemic paraphasias in spontaneous speech. It has been suggested that such patients have insufficient access to phonological information for output and cannot construct the appropriate sequence of selected phonemes for articulation. Consistent with such a proposal, we found that PW was impaired on a variety of verbal tasks that demand access to phonological representations (reading, repetition, confrontational naming and rhyme judgement); she also demonstrated poor performance on syntactic and grammatical processing tasks. However, examination of PW's repetition performance also revealed that she made semantic paraphasias and that her performance was influenced by imageability and lexical status. Her auditory–verbal short-term memory was also severely compromised. These features are consistent with 'deep dysphasia', a disorder reported in patients suffering from stroke or cerebrovascular accident, and rarely reported in the context of non-fluent progressive aphasia. PW's pattern of performance is evaluated in terms of current models of both non-fluent progressive aphasia and deep dysphasia.

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Primary diagnosis of interest

Deep dysphasia, non-fluent progressive aphasia

Author's designation of case

PW

Key theoretical issue

- Detailed exploration of the pattern of impairment in non-fluent progressive aphasia
- Detailed exploration of the pattern of impairment in deep dysphasia
- Evaluation of these impairments with reference to current interactive activation models of language function

Key words: deep dysphasia; non-fluent progressive aphasia; semantic paraphasia; phonemic paraphasia

Scan, EEG and related measures

CT, MRI, EEG

Other assessment

Detailed assessment of reading, naming, repetition, phonological processing and auditory verbal short-term memory

Lesion location

- Not present

Language

English