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Note

Progressive aphasia presenting with deep dyslexia and dysgraphia

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ABSTRACT

Primary progressive aphasia is clinically heterogeneous. We report a patient, alias Don, with a novel form of progressive aphasia, characterised by deep dyslexia and dysgraphia and dissociated access to phonological and orthographic word forms. The hallmarks of deep dyslexia and dysgraphia were present early in the course and persisted over time. Writing was initially poorer than reading, but this reversed over time. There was a lack of concordance between reading and writing errors. Don benefited from a semantic mediation strategy to learn letter sounds, involving associating letters with a country name (e.g., A = Afghanistan). Remarkably, he continued to be able to generate those phonologically complex country names when no longer able to name or sound letters. Don's performance is compatible with a traditional dual-route account of deep dyslexia and dysgraphia. The findings have potential practical implications for speech and language therapy in progressive aphasia. Moreover, they illustrate both the remarkable specificity yet clinical diversity in presentation of progressive aphasia.

1. Introduction

Primary progressive aphasia (PPA) is clinically heterogeneous. Contemporary classifications distinguish three major variants: semantic, non-fluent and logopenic (Amici et al., 2006; Gorno-Tempini et al., 2004, 2011; Grossman, 2010; Grossman and Ash, 2004; Mesulam et al., 2008). Nevertheless, not all patients conform to these prototypical syndromes. We describe a patient who presented with a striking disorder of reading and writing, characterised by deep dyslexia and dysgraphia. We have had the rare opportunity to examine the evolution of his language disorder over 9 years. His case is important because it broadens the range of possible presentations of PPA, highlighting the remarkable specificity with which language systems can be affected. It also has potential implications for therapy.

2. Case history

2.1. Initial presentation

At the age of 58 years this male University academic, pseudonym Don, noticed difficulty transcribing telephone messages

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recorded on his answering machine, and spelling 'little' words such as prepositions, conjunctions and auxiliary verbs. He also noticed problems reading hyphenated words that spanned columns. In conversational speech he experienced mild problems in word retrieval. Neither he nor his wife noticed difficulty in comprehension or in other cognitive domains. There was no change in his behaviour or personality, he had no physical symptoms and he was independent in activities of daily living. There was no relevant previous medical or family history.

Neurological examination 18 months after onset of symptoms was normal. Neuropsychological evaluation revealed a high functioning man with no abnormalities outside the language domain. Performance was above average on the Wechsler Adult Intelligence test, Recognition Memory for words and faces (Warrington, 1984) and executive tests of abstraction and set shifting (Nelson, 1976; Burgess and Shallice, 1997). Scores were at ceiling on perceptual and spatial tasks (Warrington and James, 1991). In language, conversational speech elicited no errors of articulation, phonology, semantics, morphology or syntax. He had a spoken span of 6 digits, and auditory matching span of 7 digits, as determined by the Psycholinguistic Assessments for Language in Aphasia (PALPA) test 13 (Kay et al., 1992). He scored 30/30 on PALPA tests of word and non-word repetition. Word comprehension (Manchester word-picture matching test), sentence comprehension (Bishop, 1989) and minimal pair discrimination (PALPA test 3, Kay et al., 1992) performance were also entirely normal. Category fluency was intact: he generated 23 animals in 1 min. Picture naming, irregular word reading and spoken and written spelling were within normal limits, albeit lower than expected given his educational background: on the demanding Graded naming test (McKenna and Warrington, 1983) he named 15 of 30 items. Letter fluency was reduced (23 words generated using the letters F, A and S). Despite the paucity of notable deficits, a standard spelling test (Baxter and Warrington, 1994) elicited a striking feature. He could not attempt to spell CASE when provided with the spoken context "Take your coat in case it rains", yet, when provided with the alternative context "Put your coat in the case" he wrote it effortlessly and without error. This observation raised the possibility that writing was influenced by word imageability/concreteness.

Computed tomography and subsequent magnetic resonance imaging showed atrophy of the left temporal lobe and single photon emission computed tomography (SPECT) imaging showed asymmetric reduction of tracer uptake in the left hemisphere. The clinical diagnosis was of early-stage PPA, assumed to be part of the spectrum of frontotemporal lobar degeneration (Neary et al., 1998).

2.2. Progression of illness

Over the nine-year follow-up period, speech fluency progressively diminished, reflecting difficulty accessing words, culminating in mutism. At no time were there articulatory problems. Despite presenting symptoms of dysgraphia, written communication deteriorated less rapidly than spoken, and he responded to questions in writing. Moreover, he carried a notebook containing useful words, to which he could point. Latterly, circumvention of communication difficulties through gesture became curtailed by limb apraxia. Comprehension also began to be compromised. Nevertheless, he remained independent in activities of daily living, and continued to enjoy leisure pursuits such as golf.

Informed written consent was obtained for publication of Don's data, which formed part of a broader clinical investigation of neurodegenerative disorders approved by the local Ethics committee.

3. Longitudinal assessment of single word processing

3.1. Methods

Don's reading, writing and naming skills were monitored longitudinally.

3.1.1. Reading and writing

Imageability effects were investigated using the PALPA reading (test 31) and written spelling (test 40) tests (Kay et al., 1992), which consist of high and low imageability and frequency words. Tests were re-administered at intervals over 4 years. The PALPA non-word reading (test 36) and writing (test 45) tests were also administered.

As Don reported difficulty with 'little' words, he was also asked to read and write to dictation 14 locative terms (e.g., in, under, before, between). The tasks were repeated on four occasions over a three-year period.

3.1.2. Naming and comprehension

Spoken and written naming were assessed using a 40-item picture naming task (Snowden and Neary, 2003), which includes 10 pictures of animals, 10 fruits/vegetables, 10 articles of clothing and 10 household objects. Don was asked to name each object orally and in writing. The test was administered at intervals between 2003 and 2008.

A comprehension version of the test used the same 40 item stimuli. Don was required to match a word (e.g., tiger) with one of four pictures drawn from the same semantic category (e.g., tiger, cow, elephant, lion). The task was administered twice, once with auditory and once written stimulus presentation. To examine the relationship between naming, reading and repetition, Don was asked to read aloud and to repeat object names used in the naming test.

3.2. Results

3.2.1. Reading and writing

Don showed a striking imageability effect in reading, with superior performance for high compared to low imageable words (Fig. 1a). Over time, his reading performance declined but the imageability effect persisted (test 1: $\chi^2 = 10.1$, $p = .001$; test 2: $\chi^2 = 52.4$, $p < .0001$; test 3: $\chi^2 = 51.2$, $p < .0001$; test 4: $\chi^2 = 23.2$, $p < .0001$). A significant effect of word frequency was present only on test 4 ($\chi^2 = 10.3$, $p = .001$).

Don also showed an imageability effect in writing (Fig. 1b), which persisted over time (test 1: $\chi^2 = 5.6$, $p = .02$, test 2: $\chi^2 = 15.0$, $p < .001$; test 3: $\chi^2 = 9.2$, $p = .002$; test 4: $\chi^2 = 11.9$,

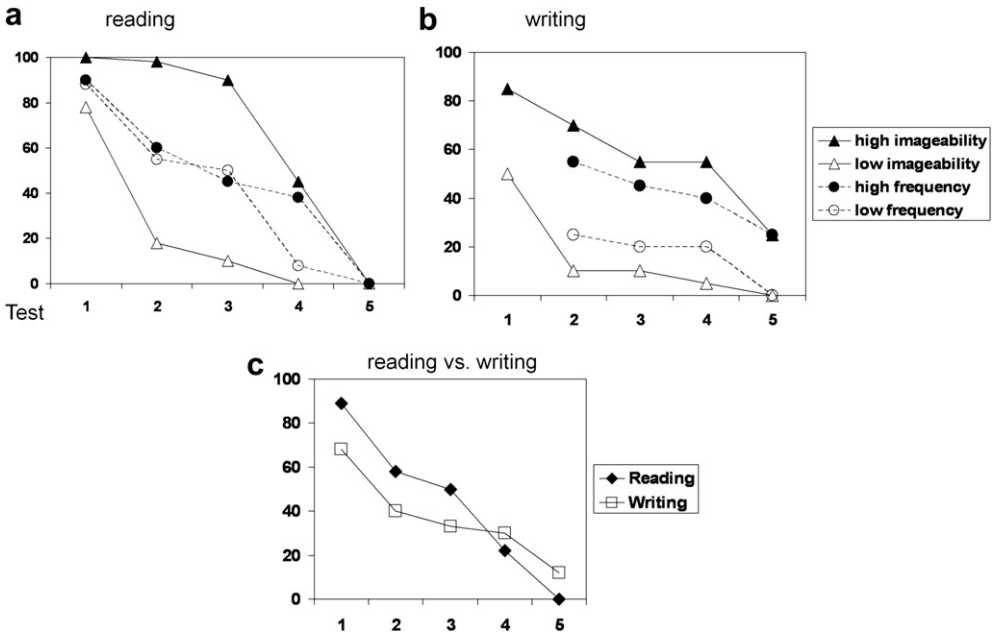

Fig. 1 – Longitudinal assessment of the effect of imageability and frequency on (a) reading and (b) writing, and (c) comparison of reading and writing for the same items.

$p = .001$; test 5: $\chi^2 = 8.5$, $p = .004$). Word frequency reached significance only at test 5 (Fisher's Exact test, $p = .02$).

Fig. 1a and b suggest superior initial performance for reading compared to writing but more precipitous decline for reading. To permit a more direct comparison between reading and writing, an item-by-item comparison was made for those 40 items common to both PALPA tasks. This confirmed the reversal from reading to writing superiority (Fig. 1c). A contingency coefficient showed an absence of correspondence between individual item reading and writing accuracy.

Reading errors were initially predominantly real word substitutions (Table 1), which included derivatives ("IDEAL" for idea) semantic substitutions ("WRY" for irony; "TELEVISION" for radio) and visually related words ("MANSION" for manner). With progression, he continued to make semantic errors although omissions accounted for an increasing percentage of responses, reflecting a total inability to attempt words.

As in reading, the largest proportion of errors in writing to dictation initially constituted real word substitutions (Table 2). These included semantic associates or synonyms (e.g., SAUCE for "gravy") and words orthographically related to the target (e.g., TUNNEL for "funnel"; MESSAGE for "marriage"), as well as occasional unrelated words (e.g., SKIPPING for "member"). He also made orthographically related non-word errors (e.g., SPINDER for "spider"; ELEVURT for "elephant"). Omissions (an inability to attempt words) accounted for an increasing proportion of responses over successive testing sessions. There was a lack of correspondence between reading and writing errors (Table 3).

Don showed considerable difficulty reading and writing non-words. Non-word reading rapidly declined from 50% correct at test 1, to 4% after 1 year and 0% thereafter. Non-word writing was 8% at test 1 and 0% at test 2. (He repeated 100% of non-words at test 1 and 75% on test 2). Errors were omissions: he responded "I don't know" and could make no attempt.

In keeping with the clinical history Don had difficulty with locative terms. Reading and writing of locatives declined at different rates. He read 100% correct at test 1, declining to 86%, 64% and 36% on subsequent tests. By contrast, he wrote only

Table 1 – PALPA imageability-frequency reading errors (% of responses)

Error type	Test 1	Test 2	Test 3	Test 4	Test 5
	01/03	04/04	01/05	09/05	06/07
Real word substitutions (semantic/visual)	9	29	20	10	0
Non-word errors (visual)	2	7	0	0	0
Omissions ("don't know")	0	6	30	67	100
Total error percentage	11	42	50	73	100

Table 2 – PALPA imageability-frequency writing errors (% of responses)

Sub-type	Test 1	Test 2	Test 3	Test 4	Test 5
Date (month/year)	01/03	09/03[a]	01/05	09/05	06/07
Real word substitutions (semantic/visual)	18	13	20	10	5
Non-word errors (orthographic)	10	30	5	5	5
Omissions ("don't know")	5	18	43	55	73
Total errors	33	61	68	70	83

a For logistical reasons test 2 for the writing task was administered at a different time from the reading task.

Table 3 – Examples of responses on reading and writing tasks

Word	Task	Patient's response			
		Test 1	Test 2	Test 3	Test 4
Idea	Read	Ideal	Why	?	?
	Write	Idea	Insight	Listening	?
Gravy	Read	Gravy	Meat	?	?
	Write	Gravy	Grovy	Sauce	Sauce
With	Read	With	With	Why	?
	Write	Was	That	For	Between
Under	Read	Under	Under	Under	Over
	Write	Below	Below	Below	Below
In front of	Read	In front of	In front of	Front of	?
	Write	Unbehind	Afront	Afront	On front inside

? = Patient responds "I don't know" and is unable to attempt.

50% correctly at test 1, with relatively s performance on repeat testing: 50%, 50% and 43%. He made semantic errors in reading and writing, with no concordance between spoken and written responses (Table 3).

3.2.2. *Naming and comprehension*

Don's naming performance approached ceiling level at test 1, but progressively declined. (Fig. 2). An item-by-item comparison, revealed superior written compared to spoken naming at each time period (test 1: $p = .02$, test 2: $p < .001$; test 3: $p < .001$, test 4: $p < .001$, test 5: $p = .001$, tests 6 and 7 not computable because of floor level spoken naming). There was a low level of concordance (correct/incorrect) between spoken and written naming responses for individual items. Contingency coefficients reached significance at test 1 ($p = .04$) and test 4 ($p = .02$), but were non-significant for other testing periods. Spoken naming performance was not influenced by semantic category or word frequency. Written naming elicited a positive effect of word frequency only at test 6 ($t = 2.4$, $p = .02$).

Semantic and associative errors (e.g., "SHOE" for sock; "RAIN JACKET" for umbrella) occurred in both spoken and written naming, but the same error never occurred in the two modalities. He made no phonological errors in spoken naming, whereas orthographic errors occurred in written naming (e.g., TIGEL for tiger; ZEDBA for zebra).

In spoken naming, omissions (inability to attempt a response) increased systematically over successive tests, accounting for 0% of responses at test 1 and 100% at test 8. By contrast, he could produce a written response even at the latest assessment.

Word comprehension performance approached ceiling levels until test 7, (Fig. 2). Subsequently, performance in the spoken and written modalities deviated, the most recent assessment revealing significantly poorer spoken than written comprehension (McNemar test $p = .007$). He also showed poorer comprehension of inanimate object terms (clothing and household objects) compared to biological category terms (animals, fruits and vegetables) in both spoken ($\chi^2 = 4.3$, $p = .04$) and written modalities ($\chi^2 = 8.6$, $p = .003$).

On early assessments Don could repeat and read words that he could not name (test 2: $p < .05$; test 3: $p < .001$). At test 4, he could repeat words that he could not read ($p = .001$) or name ($p = .002$). Differences were non-significant on subsequent tests.

Semantic errors were present in both naming and reading on early assessments, whereas they emerged in repetition only at the most recent tests. Whereas phonological errors were totally absent in spoken naming, they were occasionally present in repetition.

Omissions increased systematically over the follow-up period in all tasks.

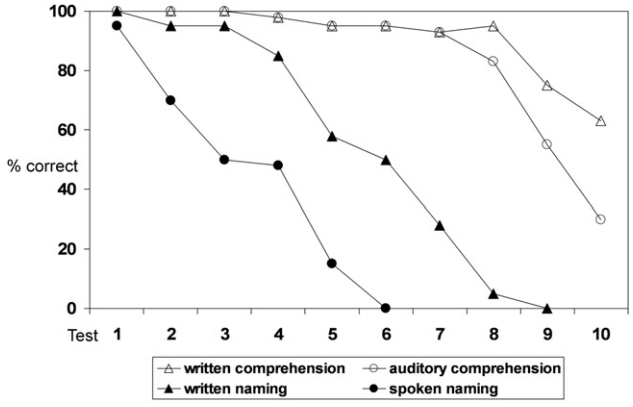

Fig. 2 – Longitudinal comparison of spoken and written naming and word comprehension

4. Speech and language therapy: semantic mediation in letter naming and sounding

Speech therapy was implemented approximately 6 months after initial assessment (before test 2). Don reported frustration at being unable to read aloud (i.e., sound out) written words. He was noted spontaneously to attempt to facilitate spoken word reading by placing the word in a meaningful, semantic context (e.g., dinner: "I eat my dinner"). As part of his language therapy, a semantic mediation strategy was implemented with the aim of facilitating access to letter names and sounds, enabling him to sound out words.

4.1. *Methods*

Don, with his therapist (JK), generated country or place names, which he learnt to associate with each written letter of the alphabet: e.g., A = Afghanistan, B = Belgium, C = Canada, E = Estonia, V = Venezuela, Z = Zambia. When he could not retrieve a written letter's sound, he was encouraged to think of the associated country name, which he would then speak aloud, thus providing him with the letter sound (e.g., V: "Venezuela/v/".) An associative strategy, by which letter names were linked to concrete words and pictures (e.g., B = bee; X = X-ray), was also adopted to teach him letter names, although imageable associates were not easy to find for all letters. Therapy focused initially on consonants, for which he had least difficulty, and then later on vowels. Don was highly motivated and practised daily.

His ability to name and sound letters was tested using the PALPA letter naming/sounding test (number 22). Performance was assessed prior to training (test 1), after 6 months, following training of consonants (test 2), after 12 months, following training of vowels (test 3), and after 2 and 3 years without further training (tests 4 and 5). His ability to generate country name associates was also monitored.

4.2. Results

At test 1, Don could name 86% of consonants and 0% of vowels. Six months later, at test 2, following training of consonants, consonant naming improved to 100%, whereas vowel naming remained at 0%. At test 3, following training of vowels, he scored 95% for consonants and 80% for vowels. Sounding data were not available at test 1. At test 2 he sounded 80% of consonants but 0% of vowels and at test 3, 64% of consonants and 80% of vowels. In view of the systematic decline in his performance for other reading tasks the improvement/maintenance of letter reading/sounding performance over a 12 months period is notable. Nevertheless, in the 2 years following completion of therapy (tests 4 and 5), letter naming and sounding performance declined to 0%.

More striking is Don's capacity to produce country names. He had no difficulty learning to produce country names in response to a letter. Moreover, despite decline in his ability to name and sound letters in the 2 years following therapy his ability to recall and reproduce the country name associated with each letter, learnt in language therapy, remained relatively robust (100% at test 3 and 92% at tests 4 and 5). At a time when he could no longer read aloud any letter names he could still generate accurately the country names linked to those letters.

5. Discussion

Don exhibits a progressive disorder of expressive language, associated with left perisylvian atrophy, in keeping with PPA. Unusually, his presenting complaints were predominantly in literacy: difficulty transcribing telephone messages, spelling prepositions and auxiliaries and reading words hyphenated across newspaper columns. Investigations elicited the hallmarks of deep dyslexia and dysgraphia: marked imageability effects, semantic errors in reading and writing and an inability to read and write non-words.

Don showed a dissociation between spoken and written performance. Despite greater initial difficulties in writing, decline was more rapid in reading. In naming, he could write down object names that he could not access orally. Perhaps the most remarkable qualitative feature of his performance was his use of a semantic strategy to assist access to word forms. He adopted this strategy spontaneously early in the course of his illness, and it was used effectively in language therapy to enable him to access letter sounds and hence sound out words. Practical benefit was relatively short-lived, lasting about 1 year. Nevertheless, he retained the ability to generate semantic associates. At a time when he could no longer name any letters he could still access the associated country name. When shown the letter A he could say Afghanistan, but not "A", the letter E "Estonia" but not "E"; the letter V "Venezuela" but not "V" and the letter Z "Zambia" but not "Z".

Deep dyslexia and deep dysgraphia have traditionally been interpreted, within the framework of the *dual-route* model (Marshall and Newcombe, 1966, 1973), as a product of damage to sublexical pathways that map orthography to phonology and vice versa. The present findings would be compatible with that view. The assumption would be that Don has particular impairment in the direct mapping between a written word and its spoken form, and to a lesser extent between a spoken word and its written form but he can access phonological and orthographic forms via the semantic system. The fact that he can produce a phonologically complex spoken word "Afghanistan" in response to the written letter A, but not the simpler and more highly-rehearsed "A", together with the absence of phonological errors in reading might reasonably argue against a primary, fundamental impairment to the phonological system, as has been reported in other patients with deep disorders (Jefferies et al., 2007). A dual *route* interpretation would be compatible with network models (Catani and Mesulam, 2008; Duffau, 2008) that have distinguished distinct neural *pathways* involved in language processing, including dorsal and ventral tracts crucial respectively for phonological and semantic aspects of language.

Deep disorders are rare in PPA. Two cases of deep dysphasia have previously been described (Majerus et al., 2001; Tree et al., 2001), but to our knowledge, no cases of deep dyslexia. Don illustrates a novel variant, providing further evidence of clinical heterogeneity within PPA.

A striking feature of some cases of PPA is relative selectivity in the disruption to psycholinguistic systems. For example, a dissociation has previously been reported between access to the spoken and written form of a word in naming tasks (Hillis et al., 2002, 2004; Holland et al., 1985; Snowden and Neary, 2003; Snowden et al., 2007). Don exemplifies such selectivity. Such dissociations are important because of their potential implications for therapy. The semantic mediation technique, used to treat Don's reading disorder, was predicated on evidence that his access to phonological forms was facilitated through semantic mediation. The use of lexical/semantic associations has been reported previously in the treatment of reading disorders following stroke (Leff and Behrmann, 2008; Ska et al., 2003), but not, to our knowledge, in PPA. Inevitably, this specific therapeutic approach would not have equal applicability to other patients with PPA. Nevertheless, Don's case exemplifies the importance of careful evaluation of patients' language disorder and person-centred language intervention in providing support for patients with PPA.

Don's language presentation does not conform to a prototypical category of PPA, as defined by current classifications (Gorno-Tempini et al., 2011): semantic, non-fluent or logopenic. Rather, he presents a novel clinical variant. His case illustrates that PPA may manifest in a remarkably pure form, as the classical syndromes of deep dyslexia and dysgraphia. The varieties of forms of PPA are yet to be fully delineated.

REFERENCES

Amici S, Gorno-Tempini ML, Ogar JM, Dronkers NF, and Miller BL. An overview on primary progressive aphasia and its variants. *Behavioural Neurology*, 17(2): 77–87, 2006.

Baxter DM and Warrington EK. Measuring dysgraphia. A graded-difficulty spelling test. *Behavioural Neurology*, 7(3-4): 107–116, 1994.

Bishop D. *TROG. Test of Reception of Grammar*. Abingdon, UK: Thomas Leach Ltd, 1989.

Burgess NW and Shallice T. *The Haylings and Brixton Tests*. Bury St Edmunds: Thames Valley test Company, 1997.

Catani M and Mesulam M. The arcuate fasciculus and the disconnection theme in language and aphasia: History and current state. *Cortex*, 44(8): 953–961, 2008.

Duffau H. The anatomo-functional connectivity of language revisited: New insights provided by electrostimulation and tractography. *Neuropsychologia*, 46(4): 927–934, 2008.

Gorno-Tempini ML, Dronkers NF, Rankin KP, Ogar JM, Phengrasamy L, Rosen HJ, et al. Cognition and anatomy of three variants of primary progressive aphasia. *Annals of Neurology*, 55(3): 335–346, 2004.

Gorno-Tempini ML, Hillis AE, Weintraub S, Kertesz A, Mendez M, Cappa SF, et al. Classification of primary progressive aphasia and its variants. *Neurology*, 76(11): 1006–1014, 2011.

Grossman M. Primary progressive aphasia: clinicopathological correlations. *Nature Reviews Neurology*, 6: 88–97, 2010.

Grossman M and Ash S. Primary progressive aphasia: a review. *Neurocase*, 10(1): 3–18, 2004.

Hillis AE, Tuffiash E, and Caramazza A. Modality specific deterioration in oral naming of verbs. *Journal of Cognitive Neuroscience*, 14(7): 1099–1108, 2002.

Hillis AE, Oh S, and Ken L. Deterioration of naming nouns versus verbs in primary progressive aphasia. *Annals of Neurology*, 55(2): 268–275, 2004.

Holland AL, McBurney DH, Moossy J, and Reinmuth OM. The dissolution of language in Pick's disease with neurofibrillary tangles: A case study. *Brain and Language*, 24(1): 36–58, 1985.

Jefferies E, Sage K, and Lambon Ralph MA. Do deep dyslexia, dysphasia and dysgraphia share a common phonological impairment? *Neuropsychologia*, 45(7): 1553–1570, 2007.

Kay J, Lesser R, and Coltheart M. *PALPA: Psycholinguistic Assessments of Language Processing in Aphasia*. Hove: Lawrence Erlbaum Associates, 1992.

Leff AP and Behrmann M. Treatment of reading impairment after stroke. *Current Opinion in Neurology*, 21(6): 644–648, 2008.

Majerus S, Lekeku F, Van der Linden M, and Salmon E. Deep dysphasia: Further evidence on the relationship between phonological short-term memory and language processing impairments. *Cognitive Neuropsychology*, 18(5): 385–410, 2001.

Marshall JC and Newcombe F. Syntactic and semantic errors in paralexia. *Neuropsychologia*, 4(2): 169–176, 1966.

Marshall JC and Newcombe F. Patterns of paralexia: A psycholinguistic approach. *Journal of Psycholinguistic Research*, 2(3): 175–199, 1973.

McKenna P and Warrington EK. *Graded Naming Test*. Windsor, UK: NFER-Nelson, 1983.

Mesulam M-M, Wicklund A, Johnson N, Rogalski E, Leger GC, Rademaker A, et al. Alzheimer and frontotemporal pathology in subsets of primary progressive aphasia. *Annals of Neurology*, 63(6): 709–719, 2008.

Neary D, Snowden JS, Gustafson L, Passant U, Stuss D, Black S, et al. Frontotemporal lobar degeneration. A consensus on clinical diagnostic criteria. *Neurology*, 51(6): 1546–1554, 1998.

Nelson HE. A modified card sorting test sensitive to frontal lobe defects. *Cortex*, 12(4): 313–324, 1976.

Ska B, Garneau-Beaumont D, Chesneau S, and Damien B. Diagnosis and rehabilitation attempt of a patient with acquired deep dyslexia. *Brain and Cognition*, 53(2): 359–363, 2003.

Snowden JS and Neary D. Progressive anomia with preserved oral spelling and automatic speech. *Neurocase*, 9(1): 27–43, 2003.

Snowden JS, Pickering-Brown SM, Du Plessis D, Mackenzie IRA, Varma A, Mann DMA, et al. Progressive anomia revisited: Focal degeneration associated with progranulin gene mutation. *Neurocase*, 13(5): 366–377, 2007.

Tree JJ, Perfect TJ, Hirsh KW, and Copstick S. Deep dysphasic performance in nonfluent progressive aphasia. *Neurocase*, 7(6): 473–488, 2001.

Warrington EK. *Recognition Memory Test*. Windsor (UK): NFER-Nelson, 1984.

Warrington EK and James M. *The Visual Object and Space Perception Battery*. Bury St Edmunds (UK): Thames Valley Test Company, 1991.