Introduction

Progressive decline of language function was described more than a century ago, but the focus on the clinical characteristics, underlying pathology and disease processes associated with primary progressive aphasia (PPA) has emerged during the past three decades. There is consensus that PPA is not a single entity, but rather reflects a heterogenous group of language-impairment profiles resulting from a number of neurodegerative diseases. Three PPA subtypes are currently recognized reflecting characteristic language profiles associated with underlying neuropathologies that variously disrupt different functional components of the left hemisphere language network. The subtypes include nonfluent/agrammatic, semantic, and logopenic variants. Regardless of the variant, word retrieval problems are ubiquitous, and anomia is the most common initial complaint of individuals with PPA. Therefore, treatments to improve or sustain naming skills are particularly relevant for this population. A number of studies have investigated effects of naming treatment for individuals with PPA, and most indicate at least some relearning and improvement in lexical retrieval. In the present study, we examine the therapeutic value of a lexical retrieval treatment that promotes strategic engagement of residual cognitive abilities as a means to maximize performance. This treatment approach is particularly relevant to the heterogenous presentation of language impairments associated with PPA because it is designed to variously engage residual semantics, phonology, and orthography.

Methods

Participants

Participant 1 (P1) was a 69 year-old, right-handed woman with a Master's degree who reported a history of language decline and a working diagnosis of PPA was given a year prior to this study (no subtype suggested). She had a profile consistent with anomic aphasia with a significant naming impairment (See Table 1). Clinical CT and PET scans showed left greater than right cortical atrophy and hypoperfusion. Pathology was most notable in the left temporal lobe but extended to left parietal areas.

Participant 2 (P2) was a 73 year-old, right-handed man with 8 years of education who reported slow onset of language difficulty. Like P1, he had anomic aphasia with a marked naming impairment. Clinical MRI and PET scans revealed left-lateralized atrophy and hypoperfusion in temporal and parietal regions.

Assessment

Extensive pre-treatment assessment was conducted and summary findings are shown in Table 1. Composite scores for semantic, phonological, and orthographic processing were computed from a battery of subtests. Both participants had significant naming impairment with relatively good comprehension and semantic processing skills. Both had relatively preserved nonverbal, visual problem-solving skills, and probably had some decline in verbal working memory. They differed regarding phonological skills in that P2 had marked difficulty with tasks such as sound deletion and sound blending, as well as phonology-to-orthography correspondences. Reading and spelling skills were also markedly degraded for P2, while relatively preserved for P1. Participant 1 was able to rely on phonology to compensate for some loss of orthographic knowledge, so that she made some phonologically plausible spelling errors, such as *grose* for gross, *brum* for broom, and had a profile consistent with mild surface agraphia. P1 also demonstrated some agnosic errors on some naming tasks: e.g., "airplane" for shark, “ball” for olive. Such errors were not common for P2. In summary, differential diagnosis for the participants included logopenic and semantic variants, and their behavioral performance suggested that P1 was in the early stages of semantic dementia, whereas P2 was in the early stages of the logopenic variant of PPA.

A complementary assessment of lexical retrieval abilities was implemented using a sequence of tasks to further explore the nature of residual knowledge and the potential to use semantic, orthographic, or phonemic cues as detailed in Table 2. In response to 20 colored pictures, each lexical retrieval failure was followed by a sequence of prompts. Both participants showed the potential to generate semantic information to help cue lexical retrieval. As expected, P1 showed greater orthographic knowledge and the potential for phonemic self-cueing than P2.

Treatment

Lexical retrieval treatment was implemented for 20 items (from 60) that were not named correctly at least 2 of 3 times on baseline probes. Using the lexical retrieval cascade as the framework for treatment, the sequence moved from the most efficient cueing (spoken semantic self-cueing) to more deliberate cueing attempts from orthography or phonology. Daily homework was structured with the use of a recordable photo album that provided pictured target items and incremental cueing.

Performance was documented using a multiple baseline design for 4 sets of 5 stimulus items, with probes taken prior to each treatment session. Criterion for advancing from one set to the next was 80% or better on the confrontation naming probe.

Results

As shown in Figure 1, both participants responded well to treatment, reaching criterion on all 4 sets of words over the course of 6 weeks for P1 and 8 weeks for P2. The calculated effect sizes (*d* statistic) were 5.76 for P1 and 7.95 P2. P1 appeared to show some generalized improvement in lexical retrieval skills as indicated by an upward drift in naming of Set 3 words, whereas P1 appeared to benefit more at an item-specific level. Post-treatment assessment confirmed P1 improved lexical retrieval for untrained words from the Lexical Retrieval Cascade (15/20 named, compared to 10/20 at pre-treatment). P1 did not show improvement in the retrieval of untrained words on the confrontation naming tasks. However, both participants showed improved word retrieval on WAB picture description task (see Table 3), providing more content at a faster rate.

Following treatment, both participants appraised word retrieval skills to be “a lot better” and indicated their improvement was well worth the time and effort expended. Six-week follow-up showed good maintenance, and the durability of these changes and use of lexical retrieval strategies is still being monitored.

Discussion

The two participants in this study demonstrated progressive lexical retrieval difficulties consistent with PPA. Both responded well to a relatively short course of behavioral treatment designed to improve reliance on residual language skills to resolve instances of anomia. Naming abilities improved for trained items and in the context of a narrative task. P1 showed additional improvement on confrontation naming of untrained items as well. Most likely, this reflected her better-preserved phonological and orthographic abilities available to complement semantic self-cueing strategies. We note, however, that P1 also had the advantage of more education and was likely to have had stronger premorbid cognitive and linguistic skills than P2.

From a clinical perspective, these treatment outcomes add to the growing literature supporting the therapeutic value of behavioral intervention for lexical retrieval deficits in PPA. For individuals in the relatively early stages of language decline, as those reported here, the response to treatment was comparable in magnitude to that documented in cases of focal damage to the left hemisphere due to stroke. Both the quantifiable measures and participant perception of change supported the value of intervention at this stage.

From a cognitive neuropsychological perspective, knowledge gained from the study of individuals with PPA complements that from the study of individuals with focal damage that is not progressive in nature. Cognitive (and computational) accounts of language processing in literate adults involve the engagement of semantics, phonology, and orthography, and greater understanding of the interactive nature of these components is gained when they are variously degraded and then re-trained, regardless of the underlying etiology.

Table 1. Participant characteristics and pre-treatment performance profiles for Participants 1 and 2.

	Demographics				Language Tests		Composite Scores		
	Age	Ed	Ravens	Digit Span	WAB AQ	BNT	Semantics	Phonology	Orthography
P1	69	18	50th %ile	30th %ile	89.8	50.0%	96.7	94.4	90.0
P2	73	8	50th %ile	6th %ile	82.2	36.7%	93.8	45.0	20.8

Ravens = Ravens Coloured Progressive Matrices; WAB = Western Aphasia Battery; BNT = Boston Naming Test.

Table 2. Lexcial Retrieval Cascade.

Lexical Retrieval Cascade

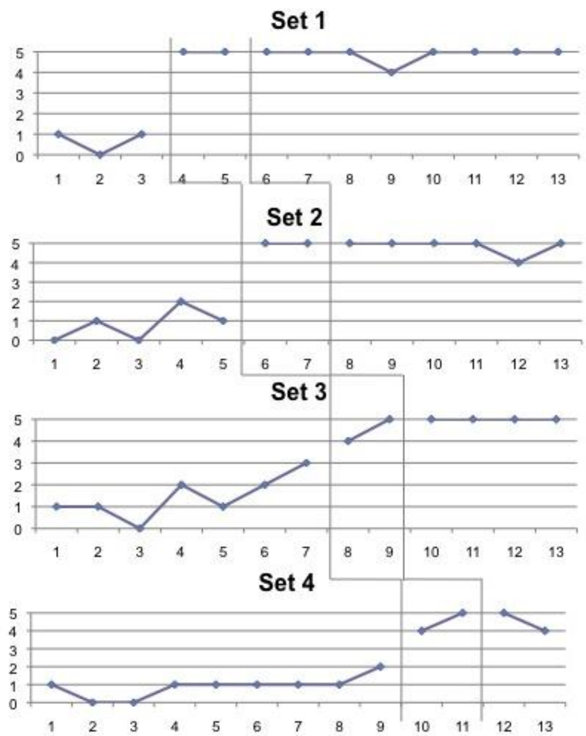
Instruction	*Task/Modality*
a. Show picture and ask, *What is this?*	Spoken naming
b. *Tell me about it.*	Semantic self-cue
c. *Can you write it? …Now can you say it?*	Orthographic self-cue
If first letter is correct. *It starts with this.*	Orthographic self-cue
d. First letter provided	
Can you say the sound for this letter?	Letter-sound correspondence
Now try to say the word.	Phonemic self-cueing
e. Present written word choices.	
Which is the correct word?	Written word recognition
If correct: *What does that say?*	Oral reading
If incorrect, point:	
This is the word. What does this say?	Oral reading
f. Prompt copying of the word.	Written copy
What does that say?	Oral reading
g. Provide phonemic cue, e.g., *It's a /d__/*	Response to phonemic cue.
h. *Repeat after me* (say word)	Spoken repetition

Table 3. Pre-Post Treatment Performance.

	Poss	Participant 1 PreTx → PostTx	Participant 2 PreTx → PostTx
AZ Lexical Retrieval Cascade	20	10 → 15	11 → 11
WAB picture description			
Informativeness (CIUs/tot words)		37.0% → 44.0%	29.0% → 42.0%
Efficiency (CIUs/min)		34.5 → 43.1	36.4 → 46.9
Boston Naming Test	60	30 → 28	22 → 24
WAB naming composite	10	8.7 → 8.1	6.9 → 7.9

Figure 1. Response to lexical retrieval treatment trained as 4 sets of 5 items each. (Multiple baseline graphs indicate number correct).

Participant 1.



Participant 2.