Primary progressive aphasia (PPA) is an acquired impairment of language caused by neurodegenerative disease affecting language regions in the brain. Unlike aphasia caused by stroke, the language impairments in PPA gradually worsen over time as atrophy spreads. Current consensus criteria for diagnosis identify three clinical variants, each of which is associated with a different pathological entity: a semantic variant, with verbal and nonverbal semantic deficits; a logopenic variant, with anomia and phonological working memory problems; and a nonfluent variant, with agrammatism and/or apraxia of speech (Gorno-Tempini et al., 2011; Gorno-Tempini et al., 2004).

Naming impairment is common to both lv and svPPA, however the nature of the deficit differs across these two syndromes. The naming impairment in lvPPA occurs as a result of damage to the phonological system, likely affecting phonological assembly or retrieval. SvPPA, on the other hand, is characterized by dramatically impaired naming in the context of a generalized semantic deficit affecting both linguistic and non-linguistic domains. Thus, anomia in svPPA is a result of damage to the semantic system, whereas anomia in lvPPA results from damage/access to phonological representations.

Relative to individuals with aphasia caused by stroke, individuals with PPA are less likely to receive treatment for speech and language deficits and there are far fewer studies examining speech-language treatment in PPA. The existing treatment literature focuses largely on remediation of naming and, of those studies, the majority of treated patients have had svPPA. There are only a couple of studies examining naming treatment in patients with lvPPA, despite the fact that naming impairment is a prominent deficit in these patients.

In this study, we administered treatment for lexical retrieval in one patient with lvPPA and one with svPPA. The two treatments were similar but not identical, with modifications in order to maximize treatment gains, given the distinct levels of cognitive-linguistic breakdown in the two syndromes.

Methods

Participants

Two individuals with PPA were identified via consensus diagnosis following comprehensive testing at the Memory and Aging Center (MAC). Diagnostic testing included neurological and neuropsychological examinations as well as extensive language testing. Structural MRI scans were also obtained. Consistent with current diagnostic guidelines, following PPA diagnosis, individuals were subsequently diagnosed by variant. One individual, presenting with relatively fluent spoken language with pauses for word-finding, occasional phonemic paraphasias, spared object knowledge, motor speech, and syntax, was diagnosed with lvPPA (hereafter, LV). MRI revealed left temporo-parietal atrophy (Figure 1). The second individual, presenting with fluent speech, marked anomia, single-word comprehension deficits, impaired object knowledge, and spared syntax and motor speech, was diagnosed with svPPA
MRI revealed left greater than right atrophy in the anterior temporal lobes.

Assessment and treatment target selection

Individuals underwent language testing pre-treatment, post-treatment, and in the case of LV, at three and six months post-treatment (Table 1). SV was not available for follow-up testing due to illness. Treatment targets were selected for LV by asking her to name color pictures of objects on three separate occasions. Items that she could not name on at least two of the three occasions were selected for treatment. Five sets of five items were selected, with each set balanced with other sets for word length, number of syllables, number of phonemes, imageability, and frequency. One set was not trained and served as a control set.

Previous research indicates that relearning of lexical targets in SV is more feasible with items for which there is some degree of residual semantic knowledge. For this reason, treatment targets for SV were selected from amongst the patient’s own possessions in her home (color photographs were taken of actual items). Items that could not be named on at least two of three consecutive occasions were eligible for treatment. Again, 20 items (in four matched sets of 5) were selected for treatment and one set of five items was selected as a control set.

Treatment methods

The in-session treatment protocol involved semantic, phonologic, and orthographic cueing. The protocol was similar across the two patients (see Table 2), but with some important differences. First, LV was trained in semantic circumlocution, whereas SV was asked to state two features only, one of which was a “personal” association, rather than a generic semantic feature. Secondly, LV was provided with first letter/sound in hopes of cueing production. Given that phonological/orthographic cueing is rarely productive in SV, our patient was provided written choices to select from (with semantically or phonologically/orthographically-similar foils). Both participants were asked to make semantic judgments regarding statements made by the clinician, then to re-state two features and provide spoken and written targets from memory. Treatment sets were trained to an 80% criterion, with one hour-long session per week.

Daily homework was administered to provide supplemental rehearsal of spoken and written targets. Homework consisted of Copy and Recall Treatment (CART; Beeson, 1999; Beeson et al., 2002; Beeson et al., 2003) with a repetition component (Beeson & Egnor, 2006). Individuals were provided a picture of the target and its written word form. They copied the written word 10 times and, after each written production, were asked to produce the spoken word form (with a model provided by a “talking” photo album for the SV participant). After ten productions, they were asked to turn over the paper and say/write the target from memory. This was repeated for each of the five targets undergoing treatment at a given time.
Results

Treatment data for trained and untrained items are shown in Figures 2 and 3. LV demonstrated a significant change in performance from pre- to post- treatment (p<.001) and pre- to three and six month follow-ups (ps<.001) for spoken production of trained items. Written production of those items also changed significantly. SV demonstrated a significant change in performance from pre- to post-treatment (p<.001) and from pre- to one month post-treatment (p<.001) for trained items. Performance was identical for both written and spoken production. Standardized tests (Table 1) revealed a gradual decline in overall language performance from pre-treatment to six months post-treatment for LV. Her confrontation naming improved slightly at post-treatment and Mini-Mental State Exam (MMSE) and Pyramids and Palm Trees Test (PPT) remained stable from pre- to post-treatment. SV showed a decline in overall language performance from pre- to post-treatment as well as a slight decline on naming and nonverbal semantic measures. MMSE also declined significantly. Both patients showed no impairment of motor speech (MSE).

Discussion

Results indicate that individuals with two variants of PPA can benefit from treatment for lexical retrieval. Successful outcomes were likely enhanced by modifications to the treatment regime based on the cognitive profile of each participant. Individuals with lvPPA can capitalize on spared semantic processing to improve phonological retrieval, whereas patients with svPPA should be encouraged to retrieve residual conceptual knowledge in conjunction with phonological and orthographic rehearsal. Our study examined simultaneous training for spoken and written naming in PPA, each of which benefited from intervention. Our LV case shows that these gains may be lasting (up to six months post-treatment). The participant with SV maintained gains up to one month post-treatment; however, information regarding maintenance beyond this time frame awaits future research.

References

Table 1. Pre- and post-treatment test results

<table>
<thead>
<tr>
<th></th>
<th>WAB-pre</th>
<th>WAB-post</th>
<th>WAB-3 mos</th>
<th>WAB-6 mos</th>
<th>BNT-pre</th>
<th>BNT-post</th>
<th>PPT-pre</th>
<th>PPT-post</th>
<th>MSE-pre</th>
<th>MSE-post</th>
<th>MMSE-pre</th>
<th>MMSE-post</th>
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</thead>
<tbody>
<tr>
<td>lvPPA</td>
<td>80</td>
<td>75.2</td>
<td>73.9</td>
<td>68.8</td>
<td>27</td>
<td>31</td>
<td>49</td>
<td>48</td>
<td>AOS-0, Dys-0</td>
<td>AOS-0, Dys-0</td>
<td>15</td>
<td>15</td>
</tr>
<tr>
<td>svPPA</td>
<td>65.9</td>
<td>60.7</td>
<td>N/A</td>
<td>N/A</td>
<td>3</td>
<td>2</td>
<td>37</td>
<td>35</td>
<td>AOS-0, Dys-0</td>
<td>AOS-0, Dys-0</td>
<td>25</td>
<td>17</td>
</tr>
</tbody>
</table>

WAB=Western Aphasia Battery, BNT=Boston Naming Test, PPT=Pyramids and Palm Trees Test (picture version), MSE=Motor Speech Evaluation (from Wertz et al., 1984), MMSE=Mini-Mental State Exam

Table 2. Treatment protocols

<table>
<thead>
<tr>
<th>LV Treatment Protocol</th>
<th>SV Treatment Protocol</th>
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<tbody>
<tr>
<td>1. A picture is placed in the center of the table in front of the participant and cards reading “is a kind of ______”, “is used for ______”, “does what?”, “has/is ______”, “is found ______”, and “reminds me of ______” are placed around the item. The participant is asked to name the item and, regardless of whether the item is named, is subsequently guided through enumeration of the semantic features of the target (group, use, action, attributes/parts, location, and association).</td>
<td>1. A target picture is shown to the participant, who is asked to retrieve one personal (e.g., my daughter gave me that) and one generic (e.g., used for sitting on) semantic feature.</td>
</tr>
<tr>
<td>2. If participant can’t name the target, written first letter and spoken first sound are provided as cues. If the item still is not named, a written and spoken model are given and repeated by the participant x3.</td>
<td>2. If participant can’t name the target, four written choices are provided (the target and three semantically and/or phonologically-related foils). Following selection of the item (if correct; if incorrect, the participant is shown the correct item), written and spoken production are elicited x3.</td>
</tr>
<tr>
<td>3. The participant is asked to make a judgment regarding the semantic plausibility of five sentences regarding the target (written and spoken presentation by the clinician). Some sentences will be semantically correct (e.g., “A feather is soft and light”), while others violate some semantic feature of the target (e.g., “A feather is hard and heavy” → violates attribute feature).</td>
<td>3. The participant is asked to make a judgment regarding the semantic plausibility of five sentences using the target word (written and spoken presentation by the clinician). Some sentences will be semantically correct (e.g., “A toaster is something used to heat bread,” while others are semantically incorrect. (e.g., “A toaster is something used to dry clothes.”)</td>
</tr>
<tr>
<td>4. The participant is asked to re-state two semantic features of the target and produce its spoken and written name.</td>
<td>4. The participant is asked to re-state two semantic features of the target and produce its spoken and written name.</td>
</tr>
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</table>
Figure 1. MRI scans for LV and SV patients

LV

SV

Figure 2 a,b. Treatment data for LV (note= untrained targets remained at zero throughout)

a.

Spoken naming of trained/untrained targets

% correct

0 10 20 30 40 50 60 70 80 90 100

Pre-Tx (Dec. 2010) Post-Tx (Jan. 2011) 3 mos Post 6 mos Post

b.

Written naming of trained/untrained targets

% correct

0 10 20 30 40 50 60 70 80 90 100

Pre-Tx (Dec. 2010) Post-Tx (Jan. 2011) 3 mos Post 6 mos Post
Figure 3. Treatment data for SV