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Examining the value of lexical retrieval treatment in primary progressive aphasia: Two positive cases

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ABSTRACT

Individuals with primary progressive aphasia (PPA) suffer a gradual decline in communication ability as a result of neurodegenerative disease. Language treatment shows promise as a means of addressing these difficulties but much remains to be learned with regard to the potential value of treatment across variants and stages of the disorder. We present two cases, one with semantic variant of PPA and the other with logopenic PPA, each of whom underwent treatment that was unique in its focus on training self-cueing strategies to engage residual language skills. Despite differing language profiles and levels of aphasia severity, each individual benefited from treatment and showed maintenance of gains as well as generalization to untrained lexical items. These cases highlight the potential for treatment to capitalize on spared cognitive and neural systems in individuals with PPA, improving current language function as well as potentially preserving targeted skills in the face of disease progression.

1. Introduction

Primary progressive aphasia (PPA) is a neurological condition wherein speech and language deteriorate as a result of neurodegenerative disease affecting areas of the brain that support communication. Three variants of PPA are now widely accepted by the clinical and research communities (Gorno-Tempini et al., 2011). These include a semantic variant (svPPA), with degradation of the semantic system that results in loss of word and object knowledge; a logopenic variant (lvPPA), with impairments in naming and repetition that are phonological in nature; and a nonfluent/agrammatic variant (nfvPPA), which is characterized by syntactic impairments and motor speech deficits.

PPA is associated with atrophy affecting the language-dominant (typically left) hemisphere to a greater extent than the non-dominant hemisphere (Gorno-Tempini et al., 2004; Grossman et al., 1996; Hodges, Patterson, Oxbury, & Funnell, 1992; Mesulam, 1982). Each of the three variants of PPA is characterized by distinct patterns of atrophy and the unique speech–language syndromes that emerge are understood to reflect the topography of damage in the brain (Gorno-Tempini et al., 2011). The multimodal semantic deficits observed in svPPA are associated with atrophy in the antero-lateral temporal lobe (left greater than right), a region widely viewed to be a cortical "hub" for word and object concepts (Hodges & Patterson, 2007; Lambon Ralph, Sage, Jones, & Mayberry, 2010). By contrast, in lvPPA, atrophy is observed in posterior perisylvian cortex in the left hemisphere, a region implicated in phonological processes critical to comprehension and production of language (Gorno-Tempini et al., 2008; Henry & Gorno-Tempini, 2010). Finally, individuals with the nonfluent variant present with atrophy in left inferior frontal regions implicated in speech production and grammar (Wilson et al., 2010a).

Lexical retrieval impairment arises in each of the three variants of PPA, however, anomia is a prominent and early feature in the semantic and logopenic variants only, with sparing of word retrieval in nfvPPA until relatively advanced stages (Gorno-Tempini et al., 2004, 2011). In svPPA, degraded semantic knowledge is thought to weaken access to intact phonological representations (Lambon Ralph, McClelland, Patterson, Galton, & Hodges, 2001). By contrast, individuals with lvPPA present with phonological deficits (diminished phonological working memory and phonological paraphasias) and largely spared semantic processing (Gorno-Tempini et al., 2008; Wilson et al., 2010b). This profile suggests that lexical retrieval in these patients may be disrupted at a post-semantic or phonological stage. As such, conceptual knowledge is intact and available, but phonological representations are insufficiently accessed or assembled, as is observed in individuals with phonological impairments in the context of aphasia due to stroke (Lambon Ralph, Moriarty, & Sage, 2002; Schwartz, Dell, Martin, Gahl, & Sobel, 2006).

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Relative to the hundreds of studies examining treatment for speech and language impairments resulting from vascular lesions, the treatment literature in PPA is rather modest. The bulk of this work has addressed naming difficulty in svPPA, with only a couple of studies examining treatment in lvPPA. In svPPA, most studies have implemented treatment protocols wherein a picture or definition was rehearsed in conjunction with the written word form to encourage co-activation of semantic and orthographic/phonological representations (Graham, Patterson, Pratt, & Hodges, 1999, 2001; Heredia, Sage, Lambon Ralph, & Berthier, 2009; Mayberry, Sage, Ehsan, & Lambon Ralph, 2011; Snowden & Neary, 2002). This approach has also been modified to include a personalized or autobiographical cue, in hopes of capitalizing on relatively spared episodic memory systems to support word retrieval (Jokel, Rochon, & Leonard, 2002, 2006; Snowden & Neary, 2002). Treatment outcomes from these studies reveal significantly improved naming for trained items, with some degree of maintenance up to 6 months post-treatment in two cases (Heredia et al., 2009; Jokel et al., 2006). Together, this work served to confirm that relearning of verbal labels in individuals with svPPA is possible. However, gains were typically item- and context-specific, suggesting that retrieval of lexical items becomes reliant on episodic memory when semantic representations are degraded. Notably, this type of learning may have unintended consequences, with residual input from a degraded semantic system resulting in training-induced semantic errors, as evidenced by over-generalization of re-learned labels to semantically-related items (Mayberry et al., 2011).

Other work with svPPA utilized more elaborated cueing hierarchies to prompt or support lexical retrieval (Bier et al., 2009; Dressel et al., 2010; Jokel & Anderson, 2012; Jokel, Rochon, & Anderson, 2010; Newhart et al., 2009). These studies, which incorporated a variety of semantic, phonological, and orthographic cues, report improvements in naming performance, with generalization to untrained items in some cases (Jokel & Anderson, 2012; Jokel et al., 2010) and maintenance of gains for two or more months post-treatment in several studies (Dressel et al., 2010; Jokel & Anderson, 2012; Jokel et al., 2010). These findings indicate that a variety of cueing modalities may be beneficial in svPPA. In fact, a study directly comparing the effects of semantic versus phonological cueing approaches found both to have therapeutic value for a single participant with svPPA (Dressel et al., 2010). That study also reported changes in activation patterns on post-treatment fMRI, indicating that both semantic and phonological approaches resulted in training-induced functional reorganization of the language network.

Studies of naming treatment for individuals with the logopenic variant of PPA (lvPPA) are more limited, with only a handful of published cases. In addition to their participant with svPPA, Newhart et al. (2009) implemented their cueing hierarchy with an individual diagnosed with lvPPA. Their treatment, which involved written naming, searching for target items in a training notebook, reading, and repetition, resulted in significant improvement for trained items and generalization to untrained words, which the authors attributed to improved access to phonological word forms. Another study examined effects of a brief but intensive treatment for lexical retrieval in an individual with lvPPA, which included semantic elaboration training and massed practice in the context of generative naming tasks (Beeson et al., 2011). The treatment resulted in significant and generalized improvements in naming that were observed up to 6 months after treatment. In addition, functional MRI documented pre- to post-treatment changes in brain activation patterns that suggested increased engagement of preserved regions of the language network after treatment.

Taken together, research suggests that implementation of appropriate behavioral intervention may result in improved language performance in PPA. As described above, various forms of naming treatment, incorporating both semantic and phonological approaches, have proven beneficial. The reports that demonstrated stronger generalizability and durability of gains typically engaged multiple central language processing components (semantics, phonology, and orthography; Beeson et al., 2011; Newhart et al., 2009; Jokel & Anderson, 2012) and two of these studies (Beeson et al., 2011; Newhart et al., 2009) were unique in training the use of strategies such as semantic and orthographic *self*-cueing, which were intended to promote generalization. In this paper, we examine a treatment protocol designed to train a sequence of self-directed cueing strategies to aid in word retrieval for both trained and untrained items.

We describe implementation and outcomes of language treatment for two individuals with PPA, one with svPPA and another with lvPPA. The two syndromes differ with regard to location of neurodegeneration and specific aspects of the language phenotype. They have in common, however, the salient feature of anomia. In treating these two individuals, we aimed to capitalize on spared cognitive and neural systems by implementing treatment that promoted the use of residual semantic, phonological, and orthographic information. The two participants were treated in different laboratories, using protocols that were similar but not identical, accommodating the unique needs of each individual and the constraints of each context. The cases are presented here as a means to illustrate treatment methods and outcomes that are promising for this population.

2. Methods

2.1. Participants and assessment measures

The two individuals who participated in this study were each evaluated with comprehensive measures of speech, language, and cognition (Table 1). In addition, they received high-resolution brain scans that were analyzed using voxel-based morphometry (VBM) to reveal the pattern of cortical atrophy relative to healthy controls (for detailed methods and VBM results, see Supplemental online material). For this report, we focus on the behavioral characteristics and imaging findings that served to confirm the progressive aphasia profile and establish language performance prior to the implementation of treatment.

2.1.1. Case 1: semantic variant (SV)

SV was a 60-year-old left-handed man who reported a year-and-a-half history of language difficulty. A doctor of veterinary medicine, he had 20 years of education and was still working at the time of the initial evaluation. SV was in the process of arranging his retirement due to increasing difficulty with word retrieval and his growing uncertainty regarding word meanings, in particular, those related to diagnosing and treating the animals under his care. At the time of the initial evaluation at the University of Arizona, SV's primary complaint was impaired word retrieval, which he described as "when I'm talking, the next word disappears out of my brain." He also described increasing difficulty with spelling, indicating that he was an excellent speller prior to the onset of his current deficits. SV's conversational speech was fluent, with pauses for lexical retrieval, and some circumlocution during instances of anomia. Family history revealed that SV's mother had early onset dementia in her sixties.

A comprehensive neuropsychological evaluation was completed 4 months prior to SV's evaluation in our lab. The report from this evaluation indicated average intellectual functioning with a relative weakness in verbal convergent reasoning. Severely impaired confrontation naming was noted, as was mildly impaired verbal fluency. SV's performance on the neuropsychological battery also revealed low average to mildly impaired performance