Treatment for Lexical Retrieval in Primary Progressive Aphasia

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Abstract

In recent years there has been an increase in research describing the behavioral characteristics and underlying pathology of primary progressive aphasia (PPA). One of the earliest and most prominent features of PPA, particularly for the logopenic and semantic variants, is anomia, and this is often the symptom that prompts individuals with PPA to seek treatment. However, speech-language pathologists who encounter individuals with PPA on their clinical caseloads often have questions about how best to manage progressive language decline in these patients. A small body of literature suggests that treatment for anomia in PPA is indeed warranted, and that item-specific improvement can be expected for a majority of individuals receiving treatment. Although generalization and maintenance of treatment gains are variable in the face of progressive decline, there are some emerging themes as to treatment approaches and patient characteristics that may promote more generalized and relatively durable treatment outcomes. Ultimately, treatment that engages residual semantic, phonologic and orthographic skills, using both strategic training and stimulation, may be appropriate for PPA patients with mild-moderate anomia.

Primary Progressive Aphasia (PPA) is characterized by progressive decline in speech-language abilities, with a relative absence of impairment to other cognitive domains during initial stages of the syndrome (Mesulam, 1982, 2001). Three variants of PPA are widely accepted in the literature, each characterized by distinct patterns of behavior and cortical atrophy (Gorno-Tempini et al., 2011). The nonfluent/agrammatic variant is associated with agrammatism in language production, impaired syntax comprehension and slow, halting speech, often with features of speech apraxia. Semantic PPA is characterized by loss of object knowledge, significant anomia, and fluent but relatively empty speech; and logopenic PPA is characterized by lexical retrieval deficits and impaired repetition, but relatively intact comprehension of single words and syntax.

While the exact incidence and prevalence of PPA are not known, Mesulam (2001) estimated that as many as 20% of all individuals with dementia have PPA. Recent data from The Aging, Demographics, and Memory Study (Plassman et al., 2007) suggest that over 3 million people in the United States aged 71 and older have dementia from varying etiologies. While this does not account for dementia onset in individuals under the age of 71, based on Mesulam’s estimate, as many as 600,000 individuals in the United States may be experiencing symptoms of primary progressive language loss.

Although patients with PPA are thought to be under-referred to speech-language pathology practices (Taylor, Kingma, Croot, & Nickles, 2009), the Aphasia Research Project at the University of Arizona has seen an increase in referrals for individuals with PPA over the past decade, and anecdotal reports from speech-language pathologists (SLPs) in our community and around the
United States indicate that they, too, are seeing more patients with a diagnosis of PPA. As the understanding and classification of PPA subtypes advances, it is possible that more PPA diagnoses are being made. Furthermore, SLPs will likely continue to see an increase in referrals for PPA cases as the general population ages and the overall incidence of dementia increases.

Despite increased attention in recent years to the nature and pathology of PPA, there remains a paucity of data regarding behavioral treatment for progressive language decline. Although SLPs may be seeing more patients with a PPA profile, there is no consensus regarding best-practice for treating primary progressive aphasia. Speech-language pathologists are skilled in treating language impairments in chronic aphasia, and it has been suggested that these skills are directly applicable to managing language impairments in PPA (McNeil & Duffy, 2001). However, PPA clearly has unique clinical characteristics, including co-occurring cognitive and motor deficits as the disease progresses, in addition to the progression of the language impairment itself. Practicing speech-language pathologists who do encounter individuals with PPA have questions about how best to manage this disorder (Taylor et al., 2009). Clinicians, second party payers, and even patients and their families, may question the utility of treatment for a disorder for which decline is certain. In short, they wonder whether treatment is “worth it” for this population.

Although there is an overall scarcity of studies addressing behavioral treatment of PPA, there is a growing body of literature suggesting that language treatment is indeed “worth it.” Several recent reviews summarizing the PPA treatment literature have found that a majority of studies have reported improvement in individual performance on treated items as compared to baseline (Beeson, & Rapcsak, 2008; Carthery-Goulart et al., 2013; Croot, Nickles, Laurence, & Manning, 2009; Henry, Jokel, Graham, Rochon, & Leonard, 2014). Henry and colleagues (2008) described treatments for anomia in semantic dementia, concluding that the literature supported evidence of new learning in this population, and that “a degree of optimism is warranted” (p. 68) regarding future behavioral treatment for anomia in semantic PPA. In their 2009 review of 13 behavioral treatment studies, Croot et al. (2009) reported that all but one of 18 total participants benefitted from treatment. Carthery-Goulart and colleagues (2013) described 39 behavioral treatment studies in their recent review, all of which reported positive outcomes. Even more recently, Jokel et al. (2014) reviewed the treatment literature for lexical impairment in PPA. They concluded that “treatment gains are very promising, especially in view of the progressive nature of the disease” (p. 12).

**Lexical Retrieval Treatments in PPA**

Because one of the earliest and most prominent features of PPA is anomia, lexical retrieval difficulties are often the symptom that leads individuals to look for a diagnosis or seek treatment. Accordingly, lexical retrieval treatments are the most widely represented treatment approaches described in the literature (Carthey-Goulart et al., 2013; Jokel et al., 2014). Approaches addressing naming in semantic PPA make up the majority of the treatment studies, likely because anomia tends to be particularly severe in this population. Furthermore, the semantic variant of PPA has historically been the most consistently defined in the literature (Gorno-Tempini et al., 2011). Lexical retrieval difficulties are also prominent in logopenic PPA, but very few treatments for this variant have been described (Beeson et al., 2011; Henry et al., 2013a; Newhart et al., 2009). This may be due to the fact that logopenic PPA is a relatively new diagnostic classification (Gorno-Tempini et al., 2004, 2011). There have also been a few studies aimed at treating lexical retrieval in nonfluent/agrammatic PPA (Jokel, Cupit, Rochon, & Leonard, 2006; Jokel, Cupit, Rochon, & Leonard, 2009; Marcotte & Ansalado, 2010). However, anomia tends to emerge later in this population, and may never be as prominent a deficit as impairments of syntax or motor speech, and thus not as likely a treatment target (Gorno Tempini et al., 2004, 2011).

Because spared and impaired language processes differ with each PPA variant, so does the underlying cause of anomia in each group. For that reason, it is helpful to first consider lexical retrieval treatment in the context of each PPA variant, and then to discuss overall themes that emerge based on the treatment literature as a whole.
Treatment for Lexical Retrieval in Semantic PPA

Anomia in semantic PPA is attributed to degradation of underlying conceptual knowledge, whereas phonological and orthographic skills are believed to be relatively preserved (Lambon Ralph, McClelland, Patterson, Galton, & Hodges, 2001). Accordingly, many of the treatments addressing lexical retrieval in semantic PPA have employed approaches designed to retrain specific concepts, incorporating treatment activities such as verbal repetition and rehearsal of picture names, often paired with the written word form (Graham, Patterson, Pratt, & Heredia, Sage, Lambon Ralph, & Berthier, 2009; Hodges, 1999, 2001; Mayberry, Sage, Ehsan, & Lambon Ralph, 2011; Snowden & Neary, 2002). Many of these treatments also intended to engage residual semantic representations with semantically based tasks, such as training definitions, encouraging patients to describe semantic features, or grouping stimuli by semantic categories (Bier et al., 2009; Graham et al., 1999, 2001; Robinson, Druks, Hodges, & Garrard, 2009). Such approaches have resulted in successful re-learning of targeted items; however, treatment gains were typically item specific and maintenance was variable, but tended to be poor.

In contrast to the approach of directly remediating impaired semantics, there is evidence to suggest that capitalizing on spared abilities may also have therapeutic value. For instance, phonology is thought to be relatively preserved in semantic PPA, and studies that have directly compared semantic with phonological cueing (e.g., first sound cueing, rhyme) have demonstrated that both approaches can be effective for improving naming performance (Dressel et al., 2010; Jokel & Anderson, 2010). Episodic memory is another relatively spared cognitive process in semantic PPA. Treatments that incorporate personally relevant cueing (e.g. training the patient to describe personally meaningful definitions or contexts) in order to engage episodic memory systems have also led to successful relearning of target items, and may promote maintenance (Robinson et al., 2009; Snowden & Neary, 2002). Ultimately, semantically based treatment alone seems to be effective for item specific gains, but treatment approaches that engage multiple processes (semantic, phonologic, orthographic, and episodic memory) may contribute to better generalization and maintenance of learning (Henry et al., 2013a; Newhart et al., 2009).

Another factor that seems to be important in promoting maintenance of treatment gains in semantic PPA is stimuli selection. Personally familiar items, particularly those that the patient is involved in selecting, may facilitate motivation and overall treatment success. Furthermore, practicing existing vocabulary and incorporating re-trained words into daily communication may contribute to maintenance (Jokel et al., 2014). While these points have primarily been made in the semantic PPA treatment literature, consideration of patient interests and motivation, and attention to typical communication activities, can certainly be important factors for stimuli choice in any variant of PPA.

Treatment for Lexical Retrieval in Logopenic PPA

In contrast to semantic PPA, individuals with logopenic PPA demonstrate relatively spared semantic knowledge, but phonological processes are typically degraded. Thus, individuals with logopenic PPA are able to retrieve concepts, but anoma may result from difficulty accessing or assembling phonology. However, the few word retrieval studies that have been reported for logopenic PPA do not exclusively address remediation of phonology. Instead, two of the three anoma treatments described for this group have employed relatively complex cueing hierarchies, engaging residual phonological abilities, but also capitalizing on spared semantics and orthographic knowledge (Henry et al., 2013a; Newhart et al., 2009). Newhart and colleagues (2009) described a treatment that they believed strengthened phonological word forms using a hierarchy that involved written naming of target items, searching for written items in a notebook, and reading and verbal repetition of the items. The logopenic PPA participant reported by Henry et al. (2013a) learned self-cueing for word retrieval using semantic elaboration and reliance on residual phonologic/orthographic knowledge. Beeson et al. (2011) described a semantically based treatment that also trained
self-cueing strategies for lexical retrieval in the context of semantic elaboration and generative naming tasks. All of the logopenic PPA participants in these studies demonstrated item specific gains, and generalization to untrained items and contexts was also reported. Beeson and colleagues (2011) and Henry and colleagues (2013a) also reported maintenance of treatment gains for up to 6 months in each of their logopenic PPA participants.

**Treatment for Lexical Retrieval in Nonfluent/Agrammatic PPA**

Although lexical retrieval is not typically a primary concern in individuals with nonfluent/agrammatic PPA, lexical retrieval treatment studies have been reported in this population (Jokel et al., 2006, 2009; Marcotte & Ansalado, 2010). Marcotte and Ansalado (2010) used semantic feature analysis therapy to address naming of nouns and verbs in a patient with nonfluent/agrammatic PPA. Improved naming post-treatment was noted, but generalization and maintenance were not evaluated. Jokel and colleagues (2006, 2009) used a computer-based cueing hierarchy that involved written initial letter and whole-word cueing for lexical retrieval in nonfluent/agrammatic PPA. Naming of treated items improved, but generalization of lexical retrieval did not. However, the authors felt that the patients demonstrated some generalization to untrained syntax/sentence production tasks immediately following treatment.

Interestingly, only one treatment described in the literature appears to have directly addressed remediation of syntax in nonfluent/agrammatic PPA, despite the fact that agrammatism is a primary characteristic in this variant. Schneider, Thompson, and Luring (1996) combined pictures with gestures to facilitate sentence production. Other treatments described for nonfluent/agrammatic PPA have addressed phonologic skills through syllable tapping tasks and auditory phoneme discrimination tasks (Louis et al., 2001) and speech production in the context of structured oral reading (Henry et al., 2013b). Positive outcomes have been reported in all of these studies; however, clearly more empirical work needs to be done in this area to guide SLPs treating individuals with nonfluent/agrammatic PPA.

**A Comprehensive Treatment for Anomia in PPA: The Lexical Retrieval Cascade Approach**

At this time, there is not one specific approach that is considered best practice for treatment of anomia in PPA. Many treatments, particularly for semantic dementia, rely primarily on "stimulation," that is, rehearsal of names/concepts using repetition, reading/writing, or semantic cueing. However, there is evidence from each of the PPA variants suggesting that treatments that engage spared cognitive-linguistic processes while capitalizing on residual skills in the impaired domain(s) may be appropriate as a first line of treatment for individuals with mild to moderate naming impairment. Furthermore, an approach that trains self-cueing in the face of lexical retrieval breakdowns is worthy of consideration.

The Lexical Retrieval Cascade treatment, used in the Aphasia Research Project and described by Henry and colleagues (2013a), is one such approach (see Table 1). This treatment encourages self-cueing using residual skills in semantics, orthography and phonology, and also incorporates personally relevant cueing for patients with semantic PPA when necessary. In addition to therapy activities with the clinician, homework is encouraged to provide practice of strategies, but also for additional stimulation (i.e., verbal and written repetition of target items) that can be adjusted based on an individual’s ability to produce target items within a treatment session. The participants with logopenic and semantic PPA reported by Henry et al. (2013a) had positive treatment outcomes, including successful relearning of target stimuli, as well as generalization to untrained items and maintenance for at least 3 months post-treatment. Henry et al. (2013a) also reported the participants’ personal perceptions of treatment outcomes, something that is not consistently described in the PPA literature. When asked to rate several items regarding communication abilities, from item-specific (e.g., “ability to name practiced items”) to general (e.g., “overall confidence in
communication”), both participants indicated they felt their overall communication skills were better than before treatment.

Table 1. The Lexical Retrieval Cascade cueing hierarchy and homework protocol.

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<th><strong>Lexical Retrieval Cascade</strong></th>
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| **1. Semantic Self-Cue** | Clinician presents a picture for naming: “Tell me about it.”  
- Additional prompts may include requests for specific semantic information: “What does it look like? Where can you find it? What do you use it for?”  
- For participants with degraded semantic knowledge, prompts may elicit personal experiences with the item, e.g., “When was the last time you used this? What memories do you have about this?” |
| **2. Orthographic Self-Cue** | Clinician requests written production: “Can you write it? How about the first letter?”  
- If participant has correct or partially correct written production, prompt spoken production with, “Does that help you say it?”  
- If participant is unable to write any part of the word, clinician provides the first grapheme: “Can you write any more of the word? Does this help you say it?” |
| **3. Phonemic Self-Cue** | Clinician points to first grapheme (written either by participant or clinician): “What sound does this letter make? Does that help you say the word?” |
| **4. Oral Reading** | Clinician provides the whole written word if the participant is unable to produce the spoken name or to write the word independently: “What does this say?”  
- Request repeated copy of written word (×3) |
| **5. Repetition** | Clinician provides a spoken model when participant is unable to produce the word given the above prompts. |

**Homework Protocol:**
1. **Cascade Only**  
   **Criteria:** Production of target words within tx session with self-cues.)  
   - Lexical Retrieval Cascade steps 1–3  
   - Recordable photo album to provide picture stimuli and first grapheme/phoneme cues.  
   - Target word not provided in spoken or written form, requiring participant to use cues to generate target.

2. **Cascade + Additional Stimulation**  
   **Criteria:** Production of target words within tx session only when provided spoken or written model.)  
   - Lexical Retrieval Cascade steps 1–3 + Modified Copy and Recall Treatment with Repetition (CART; Beeson & Egnor, 2006)  
   - Written copy and spoken repetition of target word × 10.  
   - Recall of spoken and written word from memory.

Note. Adapted from Henry et al. (2013a).

The Lexical Retrieval Cascade protocol has also been implemented with relative success in the Aphasia Research Project for several individuals with either semantic PPA or logopenic PPA. In an invited platform presentation at the 2013 ASHA Convention, we described preliminary treatment results for 7 individuals with mild-moderate anomia (5 with logopenic PPA and 2 with
semantic PPA) who participated in the Lexical Retrieval Cascade Treatment (Beeson & Rising, 2013). Like the majority of the patients reported in the literature thus far, all participants demonstrated improved ability to name targeted items. Furthermore, while generalization was variable, each of the participants demonstrated some degree of generalization to either untrained naming tasks, discourse, or both. When asked to provide feedback about the treatment, all of the participants indicated that they felt treatment had positive effects for both specific items and for overall communication ability and confidence. Data on maintenance has not yet been obtained from these participants, but immediate treatment outcomes were promising. The Lexical Retrieval Cascade approach incorporates many of the elements of previously described successful treatments for anemia in PPA. It also allows for adaptations based on an individual’s diagnosed PPA variant and language profile, as well as their severity level. Thus, it is a reasonable approach for clinicians to consider when addressing lexical retrieval deficits in their patients with PPA.

**Evaluating Treatment Outcomes in the Context of Language Decline**

One of the most challenging aspects of treating patients with PPA is, of course, the inevitable decline in language abilities. From the literature to date, it is difficult to determine how initial severity of deficits and time post-onset of symptoms affects response to treatment. While it is reasonable to think that a later time post-onset may be equivalent to more impaired language processes, and that individuals with a greater degree of language impairment may not respond as well to treatment, this assumption is not yet supported by the existing literature. In fact, there is considerable inconsistency in reporting time post-onset and severity levels in the treatment literature. Furthermore, because information about the onset of symptoms is typically obtained from family members or the individuals themselves, estimates of time post-onset may not be consistently reliable. Severity, on the other hand, is a clinical variable that can be described based on objective assessment. For example, there is some evidence to suggest that partially spared semantic knowledge is necessary for successful treatment outcomes in semantic PPA (Dressel et al., 2010; Snowden & Neary, 2002), thus, a patient’s performance on semantic tasks may give some indication as to how the individual may respond to treatment, regardless of how long symptoms have been present. Unfortunately, there is little consistency in the literature regarding severity ratings in PPA.

Another difficulty in treating individuals with PPA is interpreting response to treatment in the context of disease progression. That is, little or no change in treated language skills may constitute a positive outcome, compared to expected decline. This is particularly difficult to evaluate in single-subject studies, although maintenance of performance on treated skills (e.g., naming) in the face of decline in untreated skills (e.g., sentence repetition), may suggest a positive treatment effect. In a group study of PPA naming ability, Farrajota et al. (2012) compared the naming performance of two groups of individuals with PPA following a period of 11 months during which one group received speech-language treatment and the other group received no treatment. Although both groups declined in their performance on a naming task, the treatment group declined less than the untreated group. The authors suggest that this difference indicates that speech-language therapy may attenuate the progression of language deficits. Despite the fact that the authors acknowledge several limitations to this study, it does provide some evidence that an outcome of little change, or even decline, following treatment may still be considered positive in the context of a progressive disease.

**Conclusion**

Many questions remain regarding the generalization and durability of the benefits from lexical retrieval treatments in PPA, and these are questions that must be addressed in future research. However, the literature describing behavioral intervention for lexical retrieval in PPA does suggest, overall, that treatment is “worth it,” and clinicians can be confident in making this recommendation to patients and their families, and to justifying treatments to second party payers. Furthermore, restitutive approaches such as lexical retrieval treatment can be done in conjunction
with compensatory approaches (e.g., the creation of personalized communication books), or as a precursor to such interventions, as patient needs change with the progression of the disease. The choice of treatment approach and stimuli is dependent on the patient profile, but a treatment that employs a cueing hierarchy addressing residual semantic, phonologic, and orthographic skills, and that trains self-cueing, may be appropriate for PPA patients with mild-moderate anomia.

References


