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**Title:** Impairment-based interventions in primary progressive aphasia: Theoretical and clinical issues.

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**Aims:**

1. To extend the literature on impairment-based treatments in PPA by reporting treatments undertaken with two new cases, CD and AC.
2. To compare outcomes across impairment based treatments in PPA and show how they are consistent with the consolidation hypothesis of long-term memory and demonstration of new learning and forgetting in semantic dementia proposed by Graham et al<sup>(1)</sup>.
3. To make recommendations about intervention planning in PPA based on the results of our studies and other published impairment-based studies in PPA.

**Case 1:** The first study involved CD, a 49-year-old woman who presented with a 30 month history of pronoun reversal for he/she, word finding and reading and writing difficulties, and no decline in activities of daily living (ADLs). She was diagnosed with moderately severe conduction aphasia with anomia and preserved semantic-conceptual organisation. MRI and SPECT showed left temporal atrophy and hypoperfusion. Naming therapy used a crossover design with two sets of 30 line drawings matched for word frequency. CD demonstrated rapid learning but maintenance only occurred with daily rehearsal and there was no generalisation to a control set. Treatment effects returned to baseline in 5-9 weeks without rehearsal. Within 12-months of initial assessment, CD was functionally non-verbal but showed preserved associative semantics. At 22-months post-assessment, when she withdrew from therapy, CD still showed no deterioration in ADLs.

**Case 2:** The participant in the second study was RG, a 63-year-old woman who presented with mild anomic dysphasia and progressively deteriorating reading and writing. Some non-language impairments were evident but she showed no significant memory deficits or personality change. Just over a year after presentation, following further deterioration in her language, a two-phase semantic treatment was commenced to improve RG's spoken picture naming using word-picture matching tasks with semantic distractors. RG improved on treated items of high and low frequency, but spoken naming of untreated items continued to deteriorate. No generalisation to untreated items occurred. Informal assessment suggested that written naming also improved, and that improvement generalised to untreated items. Improvement was maintained for high frequency but not low-frequency items at two months post-treatment. RG has maintained ADLs for over 2 years since presentation and her associative semantic abilities have only recently declined.

### **Data supporting impairment-based intervention in PPA**

- CD and AC showed learning with practice, and forgetting when practice ceased, similar to the performance of DM<sup>(1)</sup>, and consistent with Graham and colleagues' theory of learning and forgetting in semantic dementia<sup>(1)</sup>.
- Three further published cases showed a similar pattern<sup>(1-4)</sup>; despite heterogeneity of presentation and of treatments used. This suggests that the consolidation hypothesis of long term memory may have application to other progressive aphasic syndromes in addition to semantic dementia.
- Impairment-based therapies can be effective in PPA, but in all reported cases there was limited generalisation if any, and learning was only maintained with ongoing practice, then lost in a period of 5 weeks to 3 months without rehearsal<sup>(2-4)</sup>. All individuals who improved with impairment-based therapies showed relatively unimpaired performance on delayed recall and semantic tasks<sup>(2-4)</sup>.
- All four people treated for anomia (CD, AC, GP & DM) carried out a therapy that linked the phonological form of the word with semantic elaboration<sup>(1,3)</sup>.
- One reported case (AM) who did not show new learning did not benefit from rehearsal, had comparatively poor semantics, and used a rehearsal strategy based on the initial letters of words without semantic elaboration<sup>(1)</sup>.
- Repeated practice caused distress (as well as improvement on trained items for DM<sup>(1)</sup> and CD, but did not cause distress for AC).

### **Recommendations for intervention planning in PPA**

1. Thorough standardised assessment should be completed prior to therapy to determine language and cognitive impairments, site of lesion and probably neuropathology and therefore suitability for naming intervention. Current evidence suggests that people with PPA with relatively preserved semantic organisation (eg. scoring in or near normal range on Pyramids and Palm Trees<sup>(5)</sup>) show improvement in naming therapy.
2. Clients should be motivated and able to complete therapy independently at home.
3. The client should be made aware of the likely limited outcome of the therapy trial and the possible psycho-emotional distress if marked improvement is followed by a steady decline in performance when practice is ceased.
4. Therapy should be structured to measure improvement, to determine if generalisation is occurring and to allow for retesting over a non-treatment period to assess maintenance.
5. Because there is little evidence for generalisation to naturalistic contexts, the therapy should target items that are highly functional for the client. This is likely to mean assembling the set of therapy items individually for each client, based on observation of everyday communicative interactions and/or interview with a spouse or other informant. Target items should be established with multiple baselines to establish the need for relearning, and divided into treatment and control sets on the basis of baseline performance, with both sets receiving treatment in a crossover design.
6. If the therapy is for anomia, the therapy task should involve semantic and phonological processing.
7. Impairment-based interventions should always be combined with strategies to improve activity and participation levels<sup>(6-8)</sup> and encompass supportive counselling.

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