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Research report

Bringing words back to mind – Improving word production in semantic dementia

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ABSTRACT

Patients with semantic dementia (SD) have significant impairments in naming and comprehension, but demonstrate relatively intact attention, everyday memory, and visuospatial skills. Given these preserved skills, attempts have been made to help re-build vocabulary in SD patients, with promising results. Such reports, however, are generally based upon only one or two cases and have employed variable retraining methods. It is thus unclear which elements of practice are crucial to success. Over two studies, we assessed four patients undergoing a word training program, who ranged in severity from mild to severe impairments to semantic knowledge. All four participants showed significant improvements in their ability to name trained items, with no changes in untrained items over the same time period. Improvements were evident within 3 weeks of practice, and could be established from a simple, repetitive practice of word-picture pairing, carried out at the participant's home. Strong effect sizes of the treatment were found in patients with severe deficits. Maintenance of learning was observed on some follow-up assessments, although continued practice is likely to be needed to sustain naming performance. Incorporating generation tasks into the practice may be assistive, but was not essential to success. These data support the utility of implementing simple home-practice programs even for patients with significant language deficits.

1. Introduction

Semantic dementia (SD) is a form of frontotemporal dementia characterized by marked anomia and word comprehension difficulties, arising from a slowly progressive degradation of conceptual knowledge (Hodges et al., 1992). This decline is associated with focal brain atrophy of the anterior–inferior temporal lobe, and in the vast majority of cases, an accumulation of an accumulation of TAR DNA-binding protein-43 (TDP-43) inclusions (Mion et al., 2010). The erosion of the semantic system leads to increasing communication difficulties that impacts significantly on the patient's psychological well-being (Thompson et al., 2003; Medina and Weintraub, 2007), and causes considerable caregiver distress and burden (Mioshi et al., in press, 2007). Other aspects of language processing, notably syntax, phonology and speech fluency remain relatively well preserved (Gorno-Tempini et al., 2004), as do other cognitive domains, such as attention, non-verbal aspects of

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executive function, everyday memory and visuospatial skills (Hodges et al., 1992, 1999; Hodges and Patterson, 2007).

The selectivity of the deficit in SD, coupled with the relative linguistic and neuropsychological strengths, provides a strong foundation for intervention. The presence of good phonological short-term memory has recently been shown to aid the acquisition of new phoneme sequences, with positive implications for verbal learning (Jefferies et al., 2011). Further, a small but growing body of evidence exists from single-case studies demonstrating that SD patients are able to relearn object labels after repeated exposure to the pictures or objects and the word form (Funnell, 1995; Graham et al., 2001; Jokel et al., 2002, 2006, 2010; Snowden and Neary, 2002; Henry et al., 2008; Heredia et al., 2009; Dressel et al., 2010; Senaha et al., 2010).

While positive, these studies have highlighted certain limitations regarding these improvements in naming; the best outcomes arise for items that are still well comprehended (Snowden and Neary, 2002; Jokel et al., 2006, 2010), the results may not generalize beyond the trained items, and improvements typically fade within a few months post-intervention (Croot et al., 2009). More detailed analysis of published results is difficult as few studies have used the same training method in more than one person, or reported a standard metric, such as an effect size, to allow comparison across individuals or studies. The key variables for success, and the intervention strategies most effective, are therefore yet to be identified.

To date, only one person with SD (participant "CUB") has reportedly sustained her naming improvements over a 6-month period after ceasing practice (Heredia et al., 2009). The reasons for this successful maintenance are unclear. One contributing factor may relate to the frequency and length of time spent during relearning. As noted by the authors, CUB completed a daily practice of one word list for 4 weeks, in comparison to previous studies, where intervention periods have typically been restricted to 2 or 3 weeks per word list (Graham et al., 2001; Snowden and Neary, 2002; Jokel et al., 2006; Henry et al., 2008; Dressel et al., 2010), and/or required only a few sessions per week (Jokel et al., 2007; Bier et al., 2009). While improvements in naming often occur rapidly, CUB's continued rehearsal of items over a longer period of time may have been a significant factor in extending the maintenance of her improvements. Certainly, principles relating to repetition and intensity of practice have been highlighted within animal models of successful learning and may be important for inducing experience-dependent plasticity in the brain (as outlined by Kleim and Jones, 2008). Despite this, no published studies have attempted to explore the effect of an intense practice over varying lengths of time in SD relearning.

In the past, therapist availability and expense have impeded the use of more intense forms of practice, but a growing trend exists toward interventions for aphasic patients which can be run on home computers (Jokel et al., 2006, 2010; Cherney et al., 2007; Katz, 2010; Mason et al., 2011). This approach has the additional benefit of providing training in a naturalistic setting, particularly in a condition where learning may be context dependent (Graham et al., 1999; Snowden and Neary, 2002). Successful intervention without the presence of a clinician requires a home program that maximizes effectiveness while minimizing complexity. Fortunately, the combination of elements thought to be assistive in word retrieval interventions is readily adaptable to computer programs, in delivering both phonologic and semantic inputs. Specifically, through simple computer based tasks (e.g., using Microsoft Powerpoint) pictures may be presented with both the spoken and written form of the word, and may be accompanied by a semantic description, so that the person sees, hears, and is encouraged to say the word aloud, all independent of a therapist's presence. In so doing, a person could engage in a daily, multi-modal practice over an extended period of time without significant expense or therapist time.

Another important factor for maintenance of words relates to their ongoing use – ideally through generalization of learning into everyday speech, as this mechanism allows for continued incidental practice after formal training ends. Although generalization to speech cannot fully explain CUB's preserved learning, given some words were not part of everyday conversation (e.g., "dromedary") and her conversation reportedly deteriorated significantly at the follow-up assessment, given relearning may be heavily dependent on short-term memory systems, some form of ongoing practice may be necessary to continually refresh memory of the words in order to sustain benefits (Graham et al., 1999). As yet, no studies in SD have described methods to bridge the gap between formal practice and a sustained, everyday use of words. Some attempt in stroke patients with aphasia has been described, including sessions where the patient is encouraged to use trained words in ways that simulate or approximate everyday use, while pictures and written cues are present (Hickin et al., 2007). Exercises ranged in complexity but included activities such as making shopping inventories, reminiscing or telling anecdotes about a chosen item, or engaging in conversation with the therapist regarding specific items; however, such an approach involves significant therapist input over multiple sessions. As sentence processing is often spared in SD, and related to the functional goal of carrying trained words into speech, one approach could be to include sentence generation into the formal practice. As a conversational task, this may be difficult, depending on the availability of others at home and the type of words selected for practice. A simple pencil and paper sentence generation task, however, may provide a stepping stone by encouraging participants to make use of the words beyond the learning context, in a format closer to everyday use. By also promoting a more active form of learning, recall of words may be further strengthened (as discussed in Wilson et al., 1994). Thus, in the same manner as providing a rich combination of inputs to the computer practice, the addition of a simple daily writing task, with the instruction to repeat the sentence aloud once complete, could further assist learning and maintenance of target words.

The purpose of the current study was to use a simple home-based method of word relearning to improve naming in a series of participants with SD. In particular, we sought to investigate some of the training variables which may affect maintenance of naming improvements, hypothesizing that:

(1) an intense daily practice would improve naming ability, with effect size potentially related to severity of semantic impairment;