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Research report

Phonological short-term memory in logopenic variant primary progressive aphasia and mild Alzheimer's disease



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ABSTRACT

It has been argued that individuals with logopenic variant primary progressive aphasia (lvPPA) have an impairment of the phonological loop, which is a component of the shortterm memory (STM) system. In contrast, this type of impairment is not thought to be present in mild typical Alzheimer's disease (AD). Thus, one would predict that people with lvPPA would score significantly lower than a matched AD group on tasks that require phonological STM. In the current study, an lvPPA group was compared with a mild AD group that was matched on age, education, and general cognitive functioning. For a subset of the tasks that involved pseudowords, the AD and lvPPA groups were compared to a healthy control group that was matched on age and education. The lvPPA group was more impaired than the AD group on all of the tasks that required phonological STM, including the pseudoword tasks, but there were no significant differences between these groups on tasks that required visuospatial STM. Compared to the healthy controls, the lvPPA group performed significantly worse on the repetition and reading of pseudowords, while the AD group did not differ significantly from the controls on these tasks. These findings are consistent with the hypothesis that phonological STM is impaired in lvPPA.

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1. Introduction

Logopenic variant primary progressive aphasia (lvPPA) is a clinical syndrome that involves impairment in word retrieval and repetition of phrases and sentences (Gorno-Tempini et al., 2011). Phonological speech errors may also occur. Single-word comprehension, object knowledge, motor speech, and grammar are typically spared. In a majority of cases, neuropathological and biomarker studies have associated lvPPA with an atypical presentation of Alzheimer's disease (AD) (Leyton et al., 2011; Mesulam et al., 2008, 2014; Rabinovici et al., 2008; Rohrer et al., 2010; Rohrer, Rossor, & Warren, 2012; Teichmann et al., 2013). In this atypical presentation, cortical atrophy is maximal within the left inferior





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parietal lobe and the left posterior superior temporal lobe (Gorno-Tempini et al., 2004, 2008; Josephs et al., 2013; Rohrer et al., 2010; Teichmann et al., 2013). In contrast, the typical presentation of AD involves degeneration that originates within the medial temporal lobe (Braak & Braak, 1995; Delacourte et al., 1999).

Gorno-Tempini and colleagues (Gorno-Tempini et al., 2004, 2008) have argued that the core mechanism that underlies the lvPPA syndrome is an impairment of the phonological loop, a short-term storage system in Baddeley's model of working memory (Baddeley, 1992, 2003a, 2012). According to this model, the phonological loop comprises two components: the phonological store, which holds speech sounds for 1–2 sec; and subvocal rehearsal, which can be utilized to refresh the speech sounds that are being held within the phonological store. The phonological store has been associated with left inferior parietal cortex, while subvocal rehearsal has been associated with left inferior frontal cortex (Baldo & Dronkers, 2006).

The existence of the phonological store is supported by the phonological similarity effect (Baddeley, 1966). This effect involves superior short-term recall of letter or word sequences with dissimilar phonology (e.g., *cow*, *bar*, *day*, *pit*), compared to letter or word sequences with similar phonology (e.g., *mad*, *mat*, *cap*, *cat*). The existence of subvocal rehearsal is supported by the word length effect (Baddeley, Thomson, & Buchanan, 1975). The word length effect involves superior short-term recall for word sequences that can be articulated quickly (e.g., *sum*, *wit*, *harm*, *bond*), compared to word sequences that require additional time for articulation (e.g., *association*, *considerable*, *university*, *representative*).

Consistent with the hypothesis that lvPPA involves an impaired phonological loop, individuals with lvPPA have exhibited deficits in digit, letter, and word span tasks (Crutch, Lehmann, Warren, & Rohrer, 2013; Foxe, Irish, Hodges, & Piguet, 2013; Gorno-Tempini et al., 2008; Leyton et al., 2014; Rohrer et al., 2010; Wilson et al., 2010). In this population, the phonological similarity effect has been found to be absent (Gorno-Tempini et al., 2008) or abnormal (Leyton et al., 2014), suggesting that the phonological store is impaired in lvPPA. In contrast, the word length effect is present in lvPPA, suggesting that subvocal rehearsal is relatively intact (Gorno-Tempini et al., 2008).

Unlike lvPPA, an impaired phonological loop is not thought to be present in mild typical AD (Foxe et al., 2013; Huntley & Howard, 2010; Leyton et al., 2014). Instead, performance on span tasks suggests that spatial short-term memory (STM) is impaired in mild AD (Huntley & Howard, 2010). Performance on block design, a task that involves visuospatial processing, is also lower in mild AD, compared to age-matched controls (Caccappolo-van Vliet et al., 2003; Ennok, Anni, Burk, & Linnamagi, 2014). Therefore, one would predict that people with lvPPA would score significantly lower than a matched AD group on tasks that require phonological STM, and one would predict that individuals with mild AD would perform significantly worse than a matched lvPPA group on tasks that require spatial STM. The former prediction, but not the latter prediction, has been supported by the performance of matched lvPPA and AD groups on digit and spatial span tasks (Foxe et al., 2013). Compared to the AD group, lvPPA

participants performed significantly worse on forward and backward digit span, while the two groups did not differ on forward and backward spatial span.

In addition to phonological span tasks, individuals with lvPPA perform significantly worse than controls on repetition tasks, including single-word and sentence repetition (Crutch et al., 2013; Leyton et al., 2014; Rohrer et al., 2010) and pseudoword repetition (Crutch et al., 2013). Individuals with AD have scored lower than healthy controls on sentence repetition (Leyton et al., 2014), but they have also scored higher than those with lvPPA on both word and sentence repetition (Foxe et al., 2013). It remains to be seen if this repetition advantage extends to pseudowords. Since pseudowords are novel and meaningless stimuli that cannot be semantically recoded, the repetition of these items provides a purer test of phonological STM (Baddeley, Gathercole, & Papagno, 1998; Friedman, 1996). Furthermore, compared to word repetition, pseudoword repetition is dependent on larger areas of cortex within the left superior temporal gyrus and the left inferior parietal lobe (Baldo, Katseff, & Dronkers, 2012), suggesting that pseudoword repetition places a greater demand on phonological STM.

Pseudoword reading is another task that has been associated with phonological STM (Bisiacchi, Cipolotti, & Denes, 1989; Butterworth, Campbell, & Howard, 1986; Caramazza, Basili, Koller, & Berndt, 1981). In previous studies, individuals with lvPPA have had lower pseudoword reading accuracy than controls (Brambati, Ogar, Neuhaus, Miller, & Gorno-Tempini, 2009; Rohrer et al., 2010), suggesting that phonological alexia is present in lvPPA. Some have argued that the phenomenon of phonological alexia results from impairment in the ability to convert graphemes to phonemes (e.g., Beauvois & Derouesne, 1979; Coltheart, Rastle, Perry, Langdon, & Ziegler, 2001), while others have argued that phonological alexia results from a modality-independent phonological processing deficit (e.g., Friedman, 1995; Patterson, Suzuki, & Wydell, 1996). In lvPPA, phonological alexia could result from a failure to hold the correct sequence of phonemes within STM (cf. Bisiacchi et al., 1989; Friedman, 1996).

In the current study, we tested the prediction that people with lvPPA would score significantly lower than a matched AD group on tasks that require phonological STM, but not on tasks that require visuospatial STM. We included multiple tests that require either phonological or visuospatial STM, and we compared an lvPPA group with a mild AD group that was matched on age, education, and general cognition, as measured by the Mini-Mental State Examination (MMSE; Folstein, Folstein, & McHugh, 1975). A subset of the phonological tests involved pseudoword tasks that are not widely available. For these tasks, the AD and lvPPA groups were compared to a healthy control group that was matched on age and education. It was predicted that the lvPPA group would perform significantly worse than the AD and control groups on tasks that require phonological STM, and it was predicted that the lvPPA and AD groups would not be significantly different on tasks that require visuospatial STM. In addition, it was predicted that the AD and control groups would have similar performance on the pseudoword tasks.

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