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Primary progressive dynamic aphasia and Parkinsonism: Generation, selection and sequencing deficits

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ABSTRACT

MC is the first reported case of dynamic aphasia in the context of non-fluent progressive aphasia and Parkinson's disease. MC's language profile was characterised by the hallmark propositional language impairment despite well-preserved naming, reading, repetition and comprehension skills. The severity of MC's propositional language deficit was comparable to other dynamic aphasic patients. Word and sentence generation performance was severely impaired only when many competing responses were activated by a stimulus. Thus, when a dominant response was available verbal generation was satisfactory. MC also presented with a deficit in idea generation and fluent sequencing of novel thoughts as discourse generation was extremely reduced and perseverative. In addition, non-verbal generation was impaired although dissociations emerged. MC was able to generate novel designs and gestures but his performance was highly perseverative, and his motor movement selection was abnormal, resembling a non-random pattern. MC is the first dynamic aphasic case with concurrent deficits in three mechanisms thought crucial for conceptual preparation processes; namely impaired *selection*, impaired *generation of ideas* and impaired *fluent sequencing of novel thoughts*. The findings are discussed in relation to conceptual preparation processes and their organisation, accounts of dynamic aphasia and the roles of the left inferior frontal and basal ganglia regions in conceptual preparation processes for verbal and non-verbal generation.

1. Introduction

Propositional language impairment in the context of well-preserved nominal and comprehension language skills is recognised as a distinct language output disorder known as *frontal dynamic aphasia* (Luria, 1970, 1973). The core feature of dynamic aphasia is severely reduced spontaneous speech in that speech is rarely initiated or used for self-expression. Luria (1970, p. 200) described these patients as able to answer direct questions easily but when storytelling, they complained of an "…emptiness in the head…." as if their thoughts "…stand still and don't move..". Thus, a sentence may be produced but difficulty remained in the formation of thoughts in connected novel speech. In contrast, naming, comprehension, reading and repetition skills are well-preserved. Luria identified the associated brain structures to be the left frontal lobe, anterior to Broca's area with the premotor cortex remaining intact. Since Luria, the language disorder of dynamic aphasia has been reported in patients with acquired focal left frontal lesions (e.g., Costello & Warrington, 1989; Robinson, Blair, & Cipolotti, 1998) and neurodegenerative conditions including progressive supranuclear palsy (PSP; e.g., Esmonde, Giles, Xuereb, & Hodges, 1996; Robinson, Shallice, & Cipolotti, 2006) and primary progressive aphasia variants (e.g., Robinson, Shallice, & Cipolotti, 2005; Snowden Griffiths, & Neary, 1996; Warren, Warren, Fox, & Warrington, 2003).

Luria was the first to provide a theoretical account of dynamic aphasia (Luria, 1970, 1973; Luria & Tsvetkova, 1967), although it was delineated as one form of transcortical motor aphasia (e.g., see Goldstein, 1948; Lichtheim, 1885). According to Luria, propositional speech impairments occur due to a breakdown in the translation of a plan into a linear scheme of a sentence. In this account, an original plan or intention was present but a subsequent breakdown in internal speech resulted in a failure to form the linear scheme and, thus, reduced propositional speech.

Since Luria, investigations of propositional speech failures have resulted in several alternative accounts of dynamic aphasia. These accounts assume a central rather than peripheral language disturbance and include a verbal planning impairment (Costello & Warrington, 1989), deficient formation and use of a lexical/semantic search strategy (Gold, Nadeau, Jacobs, Adair, Rothi, & Heilman, 1997), a failure to spontaneously activate lexical/semantic representations

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(Cox & Heilman, 2011) and a generalised deficit in initiating verbal and non-verbal responses (Raymer, Rowland, Haley, & Crosson, 2002). My colleagues and I investigated these theories in several in-depth case studies and were unable to sufficiently account for the pattern of dynamic aphasia. Thus, we proposed two distinct forms of dynamic aphasia due to a disturbance in one of two mechanisms involved in either selection from amongst competing verbal (conceptual) response propositions (Robinson et al., 1998, 2005) or fluent sequencing of novel thoughts (Robinson et al., 2006). These two mechanisms are functionally postulated to operate at the formulation or conceptual preparation stage of language production. There is recent support for both the selection and fluent sequencing of novel thought accounts (Bormann, Wallesch, & Blanken, 2008; Crescentini, Lunardelli, Mussoni, Zadini, & Shallice, 2008).

The two forms of dynamic aphasia were identified based on a review of the language generation profiles and associated lesions of the published cases (Robinson et al., 2006). Patients with the first form of dynamic aphasia fail to generate a word or sentence on generation tests and damage typically involves the left inferior frontal region (e.g., Costello & Warrington, 1989; Luria, 1970; Warren et al., 2003). This word and sentence generation impairment has been attributed to a language-specific *selection* deficit in several patients as the difficulty was only present when a stimulus activates many, compared with a dominant or few, response options or propositions that compete for selection (e.g., Crescentini et al., 2008; Robinson et al., 1998, 2005).

The second form of dynamic aphasia is less investigated although the deficit is most evident when generating multiple connected sentences or discourse rather than a single word/sentence (e.g., Esmonde et al., 1996; Robinson et al., 2006; Snowden et al., 1996). The discourse generation impairment has been attributed to a deficit in the *fluent sequencing of novel thought* that encompasses both idea generation and sequencing (Bormann et al., 2008; Robinson et al., 2006). Although it may be argued that there are very few new thoughts as we retrieve knowledge from memory, in the 19th Century Hughlings Jackson highlighted that a proposition lies within the realm of *voluntary* thoughts and is expressed in relation to a current context such that it is *novel* (reprint edited by Taylor, Holmes, & Walshe, 1932). Thus, it captures the idea that propositions are produced or generated at will and that they are new to a specific situation, in contrast to nominal language that is more automatic and constant across contexts. The majority of patients with this form of dynamic aphasia were diagnosed with PSP, or frontal lobe degeneration, with perseveration a feature for several PSP patients (for details see Robinson et al., 2006). In contrast to the first dynamic aphasia subtype, verbal and non-verbal generation deficits are evident and damage is more widespread and can encompass bilateral frontal and sub-cortical regions. For instance, neuropathological and radiological investigations of PSP have shown neuronal changes and loss bilaterally in the frontal lobes, basal ganglia and brain stem (Brenneis, Seppi, Schocke, Benke, Wenning, & Poewe, 2004; Jellinger, Bancher, Hauw, & Verny, 1995).

Recent evidence suggests dynamic aphasia can be underpinned by failures in more than one mechanism (Crescentini et al., 2008) or different mechanisms over time in the context of recovery of function (Bormann et al., 2008). Specifically, Crescentini et al. (2008) reported patient OTM who presented with a selection deficit and also a non-verbal generation deficit, unlike previously reported patients with a selection deficit (e.g., CH – Robinson et al., 2005). Thus, OTM's design fluency performance was reduced and perseverative, despite well-preserved motor movement generation. OTM's lesion also differed from the left inferior frontal involvement of previous dynamic aphasics with a selection deficit as he sustained left basal ganglia damage. Crescentini et al. concluded that "two deficits contribute to the overall pattern of performance… an impairment of verbal response generation in conditions of high competition [*a selection deficit*] is present along with one of novel thought generation evidenced by perseveration [in non-verbal generation] and due to either failure of inhibition or inability to generate novel content. These two deficits may be related to damage of different regions within the fronto-striatal circuits, namely, left frontal regions and basal ganglia" (p. 199). By contrast, Bormann et al. (2008) suggested the mechanisms underpinning dynamic aphasia may not be distinct but may arise along a continuum, based on their patient HK's performance as he recovered from a stroke. Initially, HK presented with a selection deficit on sentence generation tasks, although this resolved over 1 year such that only impaired discourse generation remained. These authors concluded this residual deficit was consistent with a novel thought generation deficit at the level of 'macro-planning' within conceptual preparation processes (Levelt, 1989, 1999). However, it is unclear whether discourse was initially impaired as only the sentence generation tests that identified the selection deficit were administered at the first time point (Bormann et al., 2008). Thus, dynamic aphasic patients with a selection deficit on word and sentence generation tasks have rarely been investigated on specific discourse generation tasks that require multiple connected sentences, other than complex scene description (but see Robinson et al., 2006). It remains unresolved whether the crucial mechanisms involved in conceptual preparation for language generation are distinct, or perhaps lie on a continuum, and whether these processes are specific to the language domain or encompasses verbal and non-verbal generation.

The neuroanatomical substrates of the two forms of dynamic aphasia have been suggested to be distinct. Cases with a language-specific selection deficit typically have left inferior frontal gyrus lesions (e.g., Costello & Warrington, 1989; Robinson et al., 1998, 2005; Warren et al., 2003). By contrast, cases with a domain-general deficit in fluent sequencing of novel thoughts tend to present with more widespread bilateral frontal and subcortical damage (e.g., Esmonde et al., 1996; Robinson et al., 2006; Snowden et al., 1996) and specifically the left basal ganglia for perseveration in non-verbal generation (Crescentini et al., 2008). The role of subcortical areas in language has become intensely researched in relation to the basal ganglia and Parkinson's disease (PD).

The basal ganglia have been implicated in lexical processing, speech initiation, syntax, production, comprehension and verbal perseveration (for review see Chan, Ryan, & Bever, 2013). Specifically in relation to dynamic aphasia, the basal ganglia have been thought crucial in lexical/semantic strategy formation/use (Gold et al., 1997) and perseveration of novel designs and gestures (Crescentini et al., 2008). A role for the basal ganglia has also been identified in inhibition of competing alternatives, not specific to language (Castner et al., 2007; Longworth, Keenan, Barker, Marslen-Wilson, & Tyler, 2005), as well as sequencing, selection and/or inhibition specific to language (Chan et al., 2013; Robles, Gatgnol, Capelle, Mitchell, & Duffau, 2005). For example, in a semantic priming paradigm individuals with PD maintain both dominant and subordinate meaning activation at longer inter-stimuli intervals, in contrast to selective priming for dominant meanings in healthy controls (Copland, de Zubicaray, McMahon, Wilson, Eastburn, & Chenery, 2003). Recently, Copland, Sefe, Ashley, Hudson, and Chenery (2009) suggested that PD is associated with impaired selection and suppression of competing representations, at least for lexical ambiguity meanings. Moreover, these authors suggest that the failure of the PD group to sustain facilitation of congruent meanings and inhibition of incongruent meanings over several intervening trials may underlie discourse processing deficits in PD. Thus, a link between selection, suppression of competing representations and fluent sequencing of novel thoughts in discourse has been hinted at although these have not yet been investigated in dynamic aphasia with Parkinson features.