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Motor Speech Disorders Associated with Primary Progressive Aphasia

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Abstract

Background—Primary progressive aphasia (PPA) and conditions that overlap with it can be accompanied by motor speech disorders. Recognition and understanding of motor speech disorders can contribute to a fuller clinical understanding of PPA and its management as well as its localization and underlying pathology.

Aims—To review the types of motor speech disorders that may occur with PPA, its primary variants, and its overlap syndromes (progressive supranuclear palsy syndrome, corticobasal syndrome, motor neuron disease), as well as with primary progressive apraxia of speech.

Main Contribution—The review should assist clinicians' and researchers' understanding of the relationship between motor speech disorders and PPA and its major variants. It also highlights the importance of recognizing neurodegenerative apraxia of speech as a condition that can occur with little or no evidence of aphasia.

Conclusion—Motor speech disorders can occur with PPA. Their recognition can contribute to clinical diagnosis and management of PPA and to understanding and predicting the localization and pathology associated with PPA variants and conditions that can overlap with them.

Keywords

primary progressive aphasia; motor speech disorders; dysarthrias; apraxia of speech; primary progressive apraxia of speech

Mesulam's (1982) widely recognized seminal description of six cases with “slowly progressive aphasia” included two individuals with buccofacial apraxia, one with limb apraxia, and one with constructional apraxia. Subsequent clinical descriptions and research related to primary progressive aphasia (PPA), including recent influential consensus criteria (Gorno-Tempini et al., 2011) -- hereafter referred to as the *consensus criteria* or *consensus paper* -- recognize that nonaphasic cognitive, affective, sensory, or motor deficits may be evident at the time of onset as long as aphasia is the primary initial symptom and predominant problem during examination early in the disease course. In some cases, an associated deficit may portend the eventual emergence of an alternative diagnosis, or may call into question the diagnosis of PPA itself. An appreciation of these associated deficits is therefore important for a full clinical understanding of PPA.

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Disorders of the motor aspects of speech production, often referred to as motor speech disorders, have been increasingly recognized as occurring with meaningful frequency in people with PPA, including early in the disease course (e.g., Croot, Ballard, Leyton, & Hodges, 2012; Josephs et al., 2006a, 2010; Ogar, Dronkers, Brambati, Miller, & Gorno-Tempini, 2007). Their recognition is important for several reasons, including their fairly predictable association (or lack thereof) with each of the clinical variants of PPA, their contribution to localization, their association with clinical and pathologic disorders that can overlap with PPA, and their influence on clinical management. In this paper we review the motor speech disorders that can be associated with and sometimes mistaken for PPA. We also address neurological conditions that overlap with PPA because they are very frequently accompanied by MSDs.

For the sake of consistency, in this paper we will adopt, whenever possible, the terms for the three PPA variants as proposed in the consensus paper: nonfluent/agrammatic PPA (nfvPPA, in many studies also called progressive nonfluent aphasia); semantic variant PPA (svPPA, also called semantic dementia); and logopenic variant PPA (lvPPA, also known as logopenic progressive aphasia). The abbreviations nfvPPA, svPPA, and lvPPA will be used unless further distinctions are required. We acknowledge that debate exists about these categories or the criteria for their diagnosis. One of our own concerns will be addressed in our discussion of primary progressive apraxia of speech.

Following a brief, broad overview of the major categories of motor speech disorders, we will review their relationships with each of the PPA variants. Clinical conditions that can overlap with PPA clinically and pathologically, namely progressive supranuclear palsy syndrome, corticobasal syndrome, and motor neuron disease, will also be discussed. We pay some special attention to apraxia of speech (AOS), with emphasis on primary progressive AOS (PPAOS), because research supports its recognition as an entity that is clinically separable from PPA in spite of it often being subsumed under the heading of PPA. We also briefly summarize clinical management issues for individuals with PPA and MSDs, or PPAOS with or without dysarthria.

MOTOR SPEECH DISORDERS

Motor speech disorders include speech disturbances that reflect abnormalities in nervous system structures or pathways directly involved in the sensorimotor programming, control, or execution of speech (Duffy, 2013). Broadly, they include dysarthrias and apraxia of speech (AOS).

Dysarthrias reflect problems with the neuromuscular control or execution of speech. There are several perceptually distinguishable types, each reflecting involvement of different portions of the central or peripheral nervous system. AOS reflects impairment in the planning or programming of movements for speech and lies at the interface between problems with language (aphasia) and motor control and execution (dysarthrias). From this perspective, AOS should be more strongly associated with PPA than are dysarthrias, and that indeed is the case. Among the different types of dysarthria that can occur in PPA, spastic and hypokinetic types, in our experience and that of others, occur most frequently

(Duffy, 2006; Josephs et al., 2013; Ogar et al., 2007). Table 1 summarizes the motor speech disorders, their general localization, their chief distinguishing perceptual characteristics, and their presumed pathophysiology. Table 2 summarizes the degree of association among each of the motor speech disorders and PPA and its associated conditions, as well as the underlying pathology typically associated with each disorder.

MOTOR SPEECH DISORDERS AND PRIMARY PROGRESSIVE APHASIA

The frequency and nature of the association between motor speech disorders and PPA is dependent on the specific PPA variant, as reviewed in the following sections. It is also important to recognize that some of the differences in the reported prevalence of dysarthria and AOS within PPA variants may reflect across-study differences in the stage of the underlying disease.

Semantic Variant PPA (svPPA)

Sparing of motor speech is one of four features, among which three must be present, required for a svPPA diagnosis, according to the consensus criteria. Although motor cortex and corticospinal tract degeneration can occur in people with TDP-43 type C pathology and svPPA (Josephs et al., 2013), the majority of studies have either documented preservation of motor speech or have not noted motor speech disorders or other motor system involvement in people with svPPA (Amici et al., 2007; Burrell, Kiernan, Vucic, & Hodges, 2011; Davies et al., 2005; Gallantucci et al., 2011; Gorno-Tempini et al., 2004; Josephs et al., 2010; Leyton et al., 2011; Rabinovici et al., 2008; Rogalski et al., 2011; Sajjadi, Patterson, Arnold, Watson, & Nestor, 2012). Rare possible exceptions are found in a study of 48 patients with svPPA that noted one case with dysarthria that was neither described nor specified as to type (Kertesz et al., 2010), and a recent study in which one patient meeting imaging criteria for svPPA (but whose clinical classification by the consensus criteria would have been nfvPPA) reportedly had AOS (Mesulam et al., 2013).

In summary, it is reasonable to conclude from the available data that motor speech disorders are very uncommon in people with svPPA. If a motor speech disorder is present, an alternative or additional clinical diagnosis should be considered.

Logopenic Variant PPA (lvPPA)

The picture regarding motor speech disorders is not quite so clear for lvPPA. The consensus criteria indicate that spared motor speech is one of four non-core features among which three must be present for lvPPA diagnosis, thus implying that motor speech disorders are a permissible attribute. Nonetheless, the consensus paper also states that preserved articulation and prosody help distinguish lvPPA from nfvPPA. At least some of this ambiguity may reflect the acknowledged at-least-occasional difficulty in distinguishing phonological paraphasias, one of the consensus criteria's supporting features for lvPPA diagnosis, from AOS features, which are strongly associated with the nfvPPA variant (Croot et al., 2012).

Several studies of varying numbers of patients have reported no evidence of motor speech disorders in people with lvPPA (Gallantucci et al., 2011; Graff-Radford, Duffy, Strand, & Josephs, 2012; Josephs et al., 2010; Leyton et al., 2011; Rabinovici et al., 2008; Sajjadi et

al., 2012). The absence of dysarthria seems consistent across many studies, although mild nonspeech parkinsonian features, and perhaps hypokinetic dysarthria, may be present in some cases (Graff-Radford et al., 2012; Josephs et al., 2010).

In contrast, the absence of AOS in lvPPA is not a universal finding. For example, Wilson et al. (2010) found that 2/10 patients with lvPPA had AOS, and although Amici et al. (2007) did not identify dysarthria in any of their 12 cases with lvPPA, ratings regarding AOS were mildly abnormal (specific abnormalities were not specified) for the lvPPA group as a whole. In a carefully conducted study that focused on the distinction between apraxic versus phonological errors in groups with nfvPPA versus lvPPA, Croot et al. (2012) found that all 14 patients with lvPPA had spared motor speech on basic clinical examination. Nonetheless, narrow transcription (conducted independent of clinically determined PPA variant classification) identified four lvPPA subjects (29%) who had definite (but not severe) evidence of AOS features (segment distortions or prosodic abnormalities), especially during repetition of polysyllabic words.

In summary, the available data and our own clinical experience support a conclusion that dysarthria is very uncommon in lvPPA. AOS is also uncommon but features of AOS may be present in a minority of cases. In general, when sound level errors occur in people with lvPPA, they are more likely to reflect phonological errors than errors attributable to AOS. Prominent dysarthria or AOS in a patient with a diagnosis of lvPPA should prompt consideration of an alternative or additional neurological diagnosis.

Nonfluent Variant PPA (nfvPPA)

Many studies note motor speech disorders in nfvPPA, with frequency ranging from low to high, but skewed toward high. In fact, several studies suggest that motor speech disorders, particularly AOS, are highly sensitive to nfvPPA (as opposed to lvPPA) and actually may be present more frequently or more prominently than agrammatism (Croot et al., 2012; Josephs et al., 2010; Knibb, Woollams, Hodges, & Patterson, 2009; Nestor et al., 2003). Some consider motor speech impairment to be a fundamental part of the syndrome or an important contributor to a diagnosis of nfvPPA/progressive nonfluent aphasia because motor speech disorders are not evident in svPPA or lvPPA (Nestor et al., 2003; Ogar et al., 2007; Rabinovici et al., 2008).

Dysarthria is more common in nfvPPA than in svPPA or lvPPA. In several studies by various research groups of varying numbers of people with nfvPPA or progressive nonfluent aphasia[i] that have explicitly noted the presence versus absence of dysarthria, dysarthria prevalence has ranged from about 20–50%[ii], with a median prevalence of 36% across the studies and a mean prevalence of 40% when the 119 cases across the studies are aggregated[iii] (Amici et al., 2007; Croot et al., 2012; Gorno-Tempini et al., 2004; Graff-Radford et al., 2012; Josephs et al., 2006a, 2010, 2013; ; Ogar et al., 2007; Rogalski et al., 2011). These estimates are in close agreement with our recently reported experience in which about one-third of our patients with both agrammatism and AOS have had dysarthria (Josephs et al., 2013). Type of dysarthria, when described, is most often spastic, hypokinetic[iv], or a mix of the two. These dysarthria types are commonly associated with progressive supranuclear palsy syndrome and corticobasal syndrome (discussed below),

whose underlying pathology is also strongly associated with nfvPPA; the dysarthria is usually less severe than the AOS which almost always accompanies it (Croot et al., 2012; Duffy, 2013; Josephs et al., 2006, 2010; Ogar et al., 2007).

In the consensus criteria, AOS is one of two core features of nfvPPA, one of which must be present for the diagnosis. The consensus paper also states that AOS may be the most common disturbance in nfvPPA and that "effortful speech and production errors can be the first symptoms of this variant, even before clear apraxia of speech or agrammatic errors occur" (Gorno-Tempini et al., 2011, p.1010).

AOS is very common across several studies of nfvPPA by different groups of investigators (cf. Ash et al., 2010), and considerably more common than dysarthria. In studies that note the presence versus absence of AOS (Amici et al., 2006, 2007; Croot et al., 2012; Gorno-Tempini, Murray, Rankin, & Weiner, 2004; Josephs et al., 2006a, 2010, 2013; Ogar et al., 2007; Rogalski et al., 2011; Rohrer, Rosser, & Warren, 2010; Wilson et al., 2010), its prevalence ranges from 17% to 100%, with a median prevalence of 78% across the studies and a mean prevalence of 69% when the 162 cases across the studies are aggregated. These summary estimates are lower than our own recently published experience in which 18 of our 19 (95%) PPA cases with agrammatism had AOS (Josephs et al., 2013). Taken together, the data suggest that AOS occurs in a majority-to-substantial-majority of people with nfvPPA. Its frequency in case series and its severity in individual cases may exceed that of aphasia.

PRIMARY PROGRESSIVE APRAXIA OF SPEECH (PPAOS)

Although this paper addresses the relationship of motor speech disorders to PPA, it is appropriate here to address primary progressive apraxia of speech (PPAOS), a disorder that we and others have concluded is separable from PPA even though many studies subsume it under the heading of PPA when there is no evidence of aphasia or when AOS is the predominant communication disorder. The merging of AOS under the diagnosis of PPA in several studies (e.g., Kertesz et al., 2003; Knibb et al., 2009; Ogar et al., 2007; Rohrer, Rosser, & Warren, 2010), at least in some instances, can be inappropriate or misleading, and may hide important information. Thus, for example, aggregating the cases across such studies suggests that about 20% of patients classified as nfvPPA/progressive nonfluent aphasia, or not otherwise specified PPA, have had little or no evidence of aphasia, and have had AOS as their primary or only deficit.

There is no doubt that AOS occurs with high frequency in people with nfvPPA in whom there is ample evidence of aphasia and agrammatism, as reviewed in the previous section and as implied in the consensus criteria. However, the consensus criteria are internally inconsistent on at least two counts. First, although the diagnostic criteria for PPA logically require that aphasia be present before determination of any PPA variant, it is stated that the "main language domains" include motor speech, thus failing to distinguish language from speech. This then seems to justify considering agrammatism and AOS as the two core features of nfvPPA, only one of which must be present for the diagnosis. As a result, a person with PPAOS (i.e., without agrammatism or other clear evidence of aphasia) could be classified as having nfvPPA. In our opinion, this ambiguity or inconsistency in the

consensus criteria should be addressed in any future revisions. This view is consistent with the suggestion that PPA should be differentiated from "states of pure progressive dysarthria or phonological disintegration, in which the formation rather than usage of words becomes disrupted" (Mesulam, Grossman, Hillis, Kertesz, & Weintraub, 2003, p. S12), and would recognize the potential contributions that attention to neurodegenerative AOS may make to the understanding of AOS in general (Duffy & Josephs, 2012).

If AOS never predominates or never occurs without aphasia, the importance of distinguishing it from nfvPPA might be lessened, but there is ample evidence that AOS can occur with no or little aphasia. This is explicitly evident in numerous studies that have documented the occurrence of AOS as the only or predominant manifestation of neurodegenerative disease and, therefore, did not designate the problem as PPA. For example, in a retrospective study of 80 patients with neurodegenerative AOS, 49% had no evidence of aphasia and 11% had no evidence of aphasia or dysarthria (Duffy, 2006; see also Josephs et al., 2012, 2006a). It is important to recognize, however, that dysarthria can be present in people with PPAOS. Our cumulative experience suggests that it occurs in about one-third of people with PPAOS; its type is most often spastic, hypokinetic, or mixed spastic-hypokinetic.

The clinical distinctiveness of PPAOS from PPA converges with anatomic and pathologic evidence. For example, voxel based morphometry, diffusion tensor imaging, and [18F]-fluorodeoxyglucose positron emission tomography have shown that PPAOS is associated with grey and white matter abnormalities of the superior lateral premotor cortex and supplementary motor area (Josephs et al., 2012; Josephs et al., 2013), in contrast to the more widespread prerolandic abnormalities, with inferior frontal lobe (and insular) predominance, that are associated with nfvPPA (e.g., Gorno-Tempini et al., 2004; Josephs et al., 2012; Nestor et al., 2003; Sapolsky et al., 2010). In addition, PPAOS and AOS-predominant-over-PPA are strongly associated with tau biochemistry and progressive supranuclear palsy or corticobasal degeneration pathologies (Josephs et al., 2006a), probably more so than is nfvPPA when AOS is not the predominant feature (Gorno-Tempini et al., 2006; Josephs et al., 2006b; Knibb et al., 2006). The relationship of PPAOS to progressive supranuclear palsy and corticobasal degeneration is discussed further in the next section.

PPA, MOTOR SPEECH DISORDERS, AND RELATED DISEASES

Progressive supranuclear palsy syndrome, corticobasal syndrome, and motor neuron disease can overlap clinically and pathologically with PPA and PPAOS (Lillo & Hodges, 2009; Josephs & Duffy, 2006; Josephs et al., 2006a; Josephs et al., 2005). People with any of these overlapping disorders who initially have no features of any of the others sometimes develop features of one of the others during disease progression (e.g., a patient with nfvPPA or PPAOS may eventually also develop cardinal features of progressive supranuclear palsy syndrome). The following sections briefly review these relationships.

Progressive supranuclear palsy syndrome

Progressive supranuclear syndrome was previously often referred to as progressive supranuclear palsy, a term now used mostly to refer to its histopathology. Progressive

supranuclear palsy syndrome is a levodopa unresponsive disease characterized clinically by axial rigidity, early falls, and vertical supranuclear gaze palsy, and pathologically by gliosis, neuronal loss, and abnormal deposition of the protein tau in glial and neuronal cells, mainly subcortically and in the brainstem regions (Josephs & Duffy, 2008). Reduced spontaneous speech, range of narrative expression, and word fluency (sometimes called dynamic aphasia) can be a presenting sign, perhaps reflecting impairment of cognitive processes related to planning and initiation of verbal output (Frattali & Duffy, 2005). Dysarthria and dysphagia are very common in progressive supranuclear palsy syndrome, and often are early and prominent signs. Dysarthria type is most often hypokinetic, spastic, or ataxic, often in various combinations (Duffy, 2013).

Aphasia and AOS are very uncommon in typical progressive supranuclear palsy syndrome (Frattali & Duffy, 2005). However, clinical and pathological studies have shown that some people with pathologically-confirmed progressive supranuclear palsy and tau pathology, or whose disease progression eventually includes clinical features of progressive supranuclear palsy syndrome, can present with AOS, agrammatic aphasia, or both, sometimes with little or no early evidence of typical clinical signs of progressive supranuclear palsy syndrome (Duffy, 2006; Josephs et al., 2005, 2006a; Roh et al., 2010; Rohrer et al., 2010). People with such clinical profiles are very likely to have progressive supranuclear palsy or corticobasal degeneration pathology (tauopathies), or both, particularly when motor speech difficulty is the most prominent symptom (Deramacourt et al., 2010; Gorno-Tempini et al., 2006; Josephs & Duffy, 2006).

Neuroimaging findings comparing cases with typical progressive supranuclear palsy syndrome versus PPAOS or nfvPPA/progressive nonfluent aphasia have identified differences between them (e.g., greater neocortical pathology and less prominent midbrain atrophy in nfvPPA/progressive nonfluent aphasia and more focal superior premotor cortex grey matter loss in PPAOS). However, enough similarities/overlap exist (e.g., grey matter loss in the supplementary motor area and reduced midbrain area) to suggest pathophysiological commonalities (Josephs et al., 2005; Rohrer et al., 2010; Whitwell et al., 2012).

Corticobasal Syndrome

Corticobasal syndrome was previously often referred to as corticobasal degeneration, a term now used mostly to refer to its histopathology, which the clinical syndrome does not always predict. Corticobasal syndrome is a levodopa unresponsive neurodegenerative disease typically characterized by asymmetric limb rigidity and apraxia as core signs as well as additional cortical and basal ganglia extrapyramidal signs (e.g., dystonia, myoclonus, bradykinesia), and pathologically by gliosis, neuronal loss and abnormal deposition of the protein tau in glial and neuronal cells. Corticobasal degeneration has clinical, pathological, biochemical, and genetic features that overlap with PPA and PPAOS (Boeve, Lang, & Litvan, 2003; Josephs & Duffy, 2006; Kertesz et al., 2000, 2003; Knibb et al., 2006).

Unlike progressive supranuclear palsy syndrome, in which aphasia and AOS are atypical, aphasia, AOS and dysarthria are common in corticobasal syndrome, with aphasia occurring in a majority of cases in some series, and dysarthria (usually spastic and/or hypokinetic) and

AOS in a substantial minority (30% to 40%) (Lehman Blake, Duffy, Boeve, Ahlskog, & Maraganore, 2003). Aphasia, AOS, and dysarthria can be evident early along with typical cardinal signs of corticobasal syndrome in some cases, or later in the disease course in others (Boeve et al., 2000; Frattali, Grafman, Patronas, Machlouf, & Litvan, 2000; Josephs and Duffy, 2008; Kertesz et al., 2000; Kertesz et al., 2000; Kertesz, McMonagle, & Jesso, 2011; Lehman Blake et al., 2003).

Similar to progressive supranuclear palsy syndrome, isolated PPA or PPAOS can also eventually progress to a syndrome that meets criteria for corticobasal syndrome (Amici et al., 2006; Josephs et al., 2006a; Kertesz et al., 2000; Lehman-Blake et al., 2003). And, as already noted, pathologic findings in people with corticobasal syndrome, nfvPPA and PPAOS are generally consistent with corticobasal degeneration or progressive supranuclear palsy (tauopathies) (Gorno-Tempini et al., 2006; Josephs & Duffy, 2006).

Motor Neuron Disease

Motor neuron disease, a disorder that predominantly affects upper and/or lower motor neurons, also shares clinical, genetic, and pathologic characteristics with frontotemporal dementia, including PPA. It has been estimated that 10–15% of patients with frontotemporal dementia have motor neuron disease, an important association because dementia in such cases tends to progress rapidly (Burrell et al., 2011; Josephs et al., 2006b). TDP-43 pathology is present in many people with sporadic and familial motor neuron disease, and in people with frontotemporal dementia associated with motor neuron disease, including those with progressive nonfluent aphasia (nfvPPA) (Lillo & Hodges, 2009).

In addition to a possible association of motor neuron disease with PPA, Duffy, Peach, and Strand (2007) documented seven cases with neurodegenerative AOS who received a clinical diagnosis of amyotrophic lateral sclerosis (ALS), the most common subcategory of motor neuron disease. Spastic or mixed spastic-flaccid dysarthria, typical of ALS, was present in all seven cases. AOS was predominant in four cases, and aphasia predominated by AOS was present in two cases and equivocally present in another two. Such observations suggest that neurodegenerative AOS does not preclude a diagnosis of ALS and that ALS should be a diagnostic consideration in people with neurodegenerative AOS.

MANAGEMENT OF MOTOR SPEECH DISORDERS IN PPA AND PPAOS

We will briefly provide an overview of treatment for the motor speech disorders that can accompany PPA or PPAOS. Although behavioral treatment of motor speech disorders is considered effective in general (e.g., Wambaugh, Duffy, McNeil, Robin, & Rogers, 2006; Yorkston, 1996), its efficacy for people with PPA or PPAOS has received limited attention (Duffy & McNeil, 2008; Henry, Meese, Truong, Babiak, Miller, & Gorno-Tempini, 2013). As a result, this overview is based primarily on evidence regarding management of motor speech disorders for other disease conditions (particularly other neurodegenerative diseases) or on our own clinical experience with people who have PPA or PPAOS. Recently published comprehensive overviews of motor speech disorder management can be found in Duffy (2013) and Yorkston, Miller, Strand, & Britton (2013).

Managing motor speech disorders in people with neurodegenerative disease can be challenging due to the diversity of motor speech disorder type and the inevitable worsening of the disorder. Obviously, in PPA and PPAOS both aphasia and motor speech disorders may require attention, and the management of any motor speech disorder will be strongly influenced not only by its nature and severity but also by the nature and severity of the aphasia.

Careful consideration must be given to appropriate staging of intervention. Immediate needs must be addressed and future problems anticipated so that compensatory strategies can be optimally timed (Yorkston, Miller, Strand, & Britton, 2013). Considered broadly, management goals in the early stages often focus on maintaining or improving intelligibility and comprehensibility of speech, followed by a shift to speech-plus-augmentative communication strategies in intermediate stages, and finally, in some cases, to augmentative and alternative means of communication (AAC). Treatment is most effective if family members and other caregivers are involved early on to ensure their understanding of the goals and strategies of therapy and to assess their ability to support or play an active part in prescribed speech and communication exercises and practice.

In the early stages, teaching basic strategies for maximizing comprehensibility (i.e., understandability of speech in context) sets the stage for habits that will be helpful throughout disease progression. For example, patients and families may practice: gaining listener attention before speaking; maintaining eye contact during communicative interactions; minimizing environmental noise and distractions; avoiding multitasking; and insuring that the topic of conversation is understood at the outset of conversation. Patients with adequate reading ability may benefit from reading aloud for brief intervals several times a day, a practice that can reinforce strategies that enhance intelligibility. These may include reducing speech rate by spacing each word, emphasizing appropriate stress and intonation, or simply focusing on “speaking clearly.” For those with hypokinetic dysarthria and reduced loudness, treatment strategies that emphasize loudness, such as Lee Silverman Voice Therapy (Fox, Ramig, Ciucci, Sapir, McFarland, & Farley, 2006), may be effective for improving both loudness and clarity. For individuals with PPAOS, reading aloud, repetitive practice with words and phrases that are particularly difficult, and work on lexical and phrasal stress may improve or help maintain intelligibility and naturalness of speech. A recent report of an individual with predominant AOS and nfvPPA has documented the effectiveness of structured oral reading in improving the production of multisyllabic words, with maintenance of gains one year following treatment (Henry et al., 2013). Script training, which involves repetitive practice on a well defined, limited number of personally relevant words or phrases used in specific communicative interactions, has been successful in people with stroke-induced moderately severe aphasia and people with AOS and less severe aphasia (Youmans, Youmans, & Hancock, 2011). As the motor speech disorder progresses, therapy may shift to strategies for repairing communication breakdowns and energy conservation. Eventually a low- or high-tech AAC system, such as an alphabet board, a text-to-speech system or even eye gaze speech generating devices may be introduced, first to augment speech and then as a primary mode of communication.

The major challenge in treating motor speech disorders in the context of PPA is that the strategies just reviewed must take into account the severity of the progressing aphasia relative to the effects of the motor speech disorder on functional communication. If aphasia is the major barrier, as it often is, and the motor speech disorder is not having a major impact on communicative efficiency or intelligibility, the focus should be on the aphasia and/or compensations for it. Depending on the nature and severity of the aphasia, this approach may include an eventual shift to more supported communication which involves teaching communication partners techniques (e.g., how to keep speech natural and acknowledge the affected person's competence, use of gestures, written key words, and drawing) that take advantage of cognitive and social abilities that are relatively preserved in order to facilitate social participation (Kagan, Black, Duchan & Simmons-Mackie, 2001).

Similarly, for those with PPAOS who eventually develop aphasia, therapy may shift from maximizing speech intelligibility to maintaining as much speech clarity as possible while adapting to the escalating language impairment. If speech becomes unintelligible, the choice of AAC must take into account the patient's language skill (especially for text to speech applications). Eventually, it may be necessary to adopt a total communication approach, using gesture, picture books, and supported communication. Assuming adequate upper extremity and visual control, planning and implementing AAC may be more effective, or at least more easily implemented, for people with PPAOS or for those whose AOS is the primary barrier to expressive communication (Duffy & McNeil, 2008). When AAC is appropriate, it is important to introduce it early, before it is needed and at a time when it can be most easily learned, so as to increase the probability that it can be used effectively when the need arises.

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REFERENCES

Amici S, Ogar J, Brambati M, Miller BL, Neuhaus J, Dronkers NL, Gorno-Tempini ML. Performance in specific language tasks correlates with regional volume changes in progressive aphasia. Cognitive and Behavioral Neurology. 2007; 20:203–211. [PubMed: 18091068]

Amici S, Gorno-Tempini ML, Ogar JM, Dronkers NF, Miller BL. An overview of primary progressive aphasia and its variants. Behavioral Neurology. 2006; 17:77–87. [PubMed: 16873918]

Ash S, McMillan C, Gunawardena D, Avants B, Morgan B, Khan A, Moore P, Grossman M. Speech errors in progressive non-fluent aphasia. Brain and Language. 2010; 113:13–20. [PubMed: 20074786]

Boeve BF, Lang AE, Litvan I. Corticobasal degeneration and its relationship to progressive supranuclear palsy and frontotemporal dementia. Annals of Neurology. 2003; 54:S15–S19. [PubMed: 12833363]

Boeve BF, Parisi J, Dickson D, Maraganore D, Ahlskog JE, Graff-Radford N, Petersen R. Demographic and clinical findings in 20 cases of pathologically-diagnosed corticobasal degeneration. Movement Disorders. 2000; 15(Suppl. 3):228.

Burrell JR, Kiernan MC, Vucic S, Hodges JR. Motor neuron dysfunction in frontotemporal dementia. Brain. 2011; 134:2582–2594. [PubMed: 21840887]

Clark DG, Charuvastra A, Miller BL, Shapira JS, Mendez MF. Fluent versus nonfluent primary progressive aphasia: A comparison of clincial and functioal neuroimaging features. Brain and Language. 2005; 94:54–60. [PubMed: 15896383]

Croot K, Ballard K, Leyton CE, Hodges JR. Apraxia of speech and phonological errors in the diagnosis of nonfluent/agrammatic and logopenic variants of primary progressive aphasia. Journal of Speech, Language, and Hearing Research. 2012; 55(Suppl):S1562–S1572.

Davies RR, Hodges JR, Kril JJ, Patterson K, Halliday GM, Xuereb JH. The pathological basis of semantic dementia. Brain. 2005; 128:1984–1995. [PubMed: 16000337]

Deramecourt V, Lebert F, Debachy B, Mackowiak-Cordoliani MA, Bombois S, Kerdraon O, Buée L, Pasquier F. Prediction of pathology in primary progressive language and speech disorders. Neurology. 2010; 74:42–49. [PubMed: 19940270]

Duffy, JR. Motor speech disorders: substrates, differential diagnosis, and management. 3rd ed.. Elsevier Mosby; St. Louis, MO: 2013.

Duffy JR. Apraxia of speech in degenerative neurologic disease. Aphasiology. 2006; 20:511–527.

Duffy JR, Josephs KA. The diagnosis and understanding of apraxia of speech: why including neurodegenerative etiologies may be important. Journal of Speech, Language, and Hearing Research. 2012; 55:S1518–S1522.

Duffy, JR.; McNeil, MR. Primary progressive aphasia and apraxia of speech. In: Chapey, R., editor. Language intervention strategies in aphasia and related neurogenic communication disorders. Lippincott Williams & Wilkins; Philadelphia: 2008. p. 543-563.

Duffy JR, Peach RK, Strand EA. Progressive apraxia of speech as a sign of motor neuron disease. American Journal of Speech-Language Pathology. 2007; 16:198–208. [PubMed: 17666546]

Fox C, Ramig L, Ciucci M, Sapir S, McFarland DH, Farley B. Science and practice of LSVT/LOUD: Neural plasticity-principled approach to treating individuals with Parkinson disease and other neurological disorders. Seminars in Speech and Language. 2006; 27:283–299. [PubMed: 17117354]

Frattali CM, Grafman J, Patronas N, Machlouf F, Litvan I. Language disturbances in corticobasal degeneration. Neurology. 2000; 54:990–992. [PubMed: 10691002]

Fratalli, CM.; Duffy, JR. Characterizing and assessing speech and language disturbances. In: Litvan, I., editor. Current Clinical Neurology: Atypical Parkinsonian Disorders. Humana Press Inc; Totowa, NJ: 2005.

Gallantucci S, Tartaglia MC, Wilson SM, Henry ML, Filippi M, Agosta F, Dronkers NF, Gorno-Tempini ML. White matter damage in primary progressive aphasias: a diffusion tensor tractography study. Brain. 2011; 134:3011–3029. [PubMed: 21666264]

Gorno-Tempini ML, Dronkers NF, Rankin KP, Ogar JM, Phengrasamy L, Rosen H, Miller BL. Cognition and anatomy in three variants of primary progressive aphasia. Annals of Neurology. 2004; 55:335–346. [PubMed: 14991811]

Gorno-Tempini ML, Hillis AE, Weintraub S, Kertesz A, Mendez M, Cappa SF, Grossman M. Classification of primary progressive aphasia and its variants. Neurology. 2011; 15:1006–1014. [PubMed: 21325651]

Gorno-Tempini ML, Murray RC, Rankin KP, Weiner MW. Clinical, cognitive, and anatomincal evolution from nonfluent progressive aphasia to corticobasal syndrome: a case report. Neurocase. 2004; 10:426–436. [PubMed: 15788282]

Gorno-Tempini ML, Ogar JM, Brambati SM, Wang P, Jeong JH, Rankin KP, Miller BL. Anatomical correlates of early mutism in progressive nonfluent aphasia. Neurology. 2006; 67:1849–1851. [PubMed: 16931509]

Graff-Radford J, Duffy JR, Strand EA, Josephs KA. Parkinsonian motor features distinguish the agrammatic from logopenic variant of primary progressive aphasia. Parkinsonism and Related Disorders. 2012; 18:890–892. [PubMed: 22575236]

Henry ML, Meese MV, Truong S, Babiak MC, Miller BL, Gorno-Tempini ML. Treatment for apraxia of speech in nonfluent variant primary progressive aphasia. Behavioral Neurology. 2013; 26:77–88. [PubMed: 22713405]

Josephs K, A. Boeve BF, Duffy JR, Smith GE, Knopman DS, Parisi JE, Dickson DW. Atypical progressive supranuclear palsy underlying progressive apraxia of speech and nonfluent aphasia. Neurocase. 2005; 11:283–296. [PubMed: 16093229]

Josephs KA, Duffy JR. Apraxia of speech and nonfluent aphasia: a new clinical marker for corticobasal degeneration and progressive supranuclear palsy. Current Opinion in Neurology. 2008; 21:688–692. [PubMed: 18989114]

Josephs KA, Duffy JR, Fossett T, Strand EA, Classen DO, Whitwell JL, Peller PJ. Fluorodeoxyglucose F18 Positron Emission Tomography in Progressive Apraxia of Speech and Primary Progressive Aphasia Variants. Archives of Neurology. 2010; 67:596–605. [PubMed: 20457960]

Josephs KA, Duffy JR, Strand EA, Machulda MM, Senjem ML, Lowe VJ, Whitwell JL. Syndromes dominated by apraxia of speech show distinct characteristics from agPPA. Neurology. 2013; 81:337–345. [PubMed: 23803320]

Josephs KA, Duffy JR, Strand EA, Machulda MM, Senjem ML, Master AV, Whitwell JL. Characterizing a neurodegenerative syndrome: primary progressive apraxia of speech. Brain. 2012; 135:1522–1536. [PubMed: 22382356]

Josephs KA, Duffy JR, Strand EA, Whitwell JL, Layton KF, Parisi JE, Petersen RC. Clinicopathological and imaging correlates of progressive aphasia and apraxia of speech. Brain. 2006a; 129:1385–1398. [PubMed: 16613895]

Josephs KA, Petersen RC, Knopman DS, Boeve BF, Whitwell JL, Duffy JR, Dickson DW. Clinicopathologic analysis of frontotemporal and corticobasal degenerations and PSP. Neurology. 2006b; 10:41–48. [PubMed: 16401843]

Josephs KA, Whitwell JL, Murray ME, Parisi JE, Graff-Radford NR, Knopman DS, Dickson DW. Corticospinal tract degeneration associated with TDP-43 type C pathology and semantic dementia. Brain. 2013; 136:455–470. [PubMed: 23358603]

Kagan A, Black SE, Duchan JF, Simmons-Mackie N. Training volunteers as conversation partners using “supported conversation for adults with aphasia” (SCA): A controlled trial. Journal of Speech, Language, and Hearing Research. 2001; 44:624–638.

Kertesz A, Davidson W, McCabe P, Takagi K, Munoz D. Primary progressive aphasia: diagnosis, varieties, evolution. Journal of the International Neuropsychological Society. 2003; 9:710–719. [PubMed: 12901777]

Kertesz A, Jesso S, Harciarek M, Blair M, McMonagle P. What is semantic dementia?: A cohort study of diagnostic features and clincial boundaries. Archives of Neurology. 2010; 67:483–489. [PubMed: 20385916]

Kertesz A, Martinez-Lage P, Davidson W, Munoz DG. The corticobasal degeneration syndrome overlaps progressive aphasia and frontotemporal dementia. Neurology. 2000; 55:1368–1375. [PubMed: 11087783]

Kertesz A, McMonagle P, Jesso S. Extrapyramidal syndromes in frontotemporal degeneration. Journal of Molecular Neuroscience. 2011; 45:336–342. [PubMed: 21887521]

Knibb JA, Woollams AM, Hodges JR, Patterson K. Making sense of progressive non-fluent aphasia: an analysis of conversational speech. Brain. 2009; 132:2734–2746. [PubMed: 19696033]

Knibb JA, Xuereb JH, Patterson K, Hodges JR. Clinical and pathological characterization of progressive aphasia. Annals of Neurology. 2006; 59:156–65. [PubMed: 16374817]

Kremen SA, Mendez MF, Tsai PH, Teng E. Extrapyramidal signs in the primary progressive aphasias. American Journal of Alzheimer's Disease & Other Dementias. 2011; 26:72–77.

Lehman Blake M, Duffy JR, Boeve B, Ahlskog JE, Maraganore DM. Speech and language associated with corticobasal degeneration. Journal of Medical Speech Language Pathology. 2003; 11:131–146.

Leyton CE, Villemagne VL, Savage S, Pike KE, Ballard KJ, Piguet O, Hodges JR. Subtypes of progressive aphasia: application of the international consensus criteria and validation using β-amyloid imaging. Brain. 2011; 134:3030–3043. [PubMed: 21908392]

Lillo P, Hodges JR. Frontotemporal dementia and motor neuron disease: Overlapping clinicopathological disorders. Journal of Clinical Neuroscience. 2009; 16:1131–1135. [PubMed: 19556136]

Mesulam MM. Slowly progressive aphasia without generalized dementia. Annals of Neurology. 1982; 11:592–598. [PubMed: 7114808]

Mesulam MM, Grossman M, Hillis A, Kertesz A, Weintraub S. The core and halo of primary progressive aphasia and semantic dementia. Annals of Neurology. 2003; 54:S11–S14. [PubMed: 12833362]

Mesulam MM, Wieneke C, Hurley R, Rademaker A, Thompson CK, Weintraub S, Rogalski EJ. Word and objects at the tip of the left temporal lobe in primary progressive aphasia. Brain. 2013; 136:601–618. [PubMed: 23361063]

Nestor PJ, Graham NL, Fryer TD, Williams GB, Patterson K, Hodges JR. Progressive non-fluent aphasia is associated with hypometabolism centered on the left anterior insula. Brain. 2003; 126:2406–2418. [PubMed: 12902311]

Ogar JM, Dronkers NF, Brambati SM, Miller BL, Gorno-Tempini ML. Progressive nonfluent aphasia and its characteristic motor speech deficits. Alzheimer Disease & Associated Disorders. 2007; 21(4):S23–S29. [PubMed: 18090419]

Rabinovici GD, Jagust WJ, Furst AJ, Ogar JM, Racine CA, Mormino EC, Gorno-Tempini ML. Aβ amyloid and glucose metabolism in three variants of primary progressive aphasia. Annals of Neurology. 2008; 64:388–401. [PubMed: 18991338]

Rogalski E, Cobia D, Harrison TM, Wieneke C, Weintraub S, Mesulam MM. Progression of language decline and cortical atrophy in subtypes of primary progressive aphasia. Neurology. 2011; 76:1804–1810. [PubMed: 21606451]

Roh JH, Suh MK, Kim EJ, Go SM, Na DL, Seo AW. Glucose metabolism in progressive nonfluent aphasia with and without parkinsonism. Neurology. 2010; 75:1022–1024. [PubMed: 20837971]

Rohrer JD, Paviour D, Bronstein AM, O'Sullivan SS, Lees A, Warren JD. Progressive supranuclear palsy syndrome presenting as progressive nonfluent aphasia: a neuropsycholgical and neuroimaging analysis. Movement Disorders. 2010; 25:179–188. [PubMed: 20077483]

Rohrer JD, Rossor MN, Warren JD. Syndromes of nonfluent primary progressive aphasia: a clinical and neurolinguistic analysis. Neurology. 2010; 75:603–10. [PubMed: 20713949]

Sajjadi SA, Patterson K, Arnold RJ, Watson PC, Nestor PJ. Primary progressive aphasia: A tale of two syndromes and the rest. Neurology. 2012; 78:1670–1677. [PubMed: 22573633]

Sapolsky D, Bakkour A, Negeira A, Nalipinski P, Weintraub S, Mesulam MM, Dickerson BC. Cortical neuroanatomic correlates of symptom severity in primary progressive aphasia. Neurology. 2010; 75:358–366. [PubMed: 20660866]

Wambaugh JL, Duffy JR, McNeil MR, Robin DR, Rogers MA. Treatment guidelines for acquired apraxia of speech: a synthesis and evaluation of the evidence. Journal of Medical Speech Language Pathology. 2006; 14:xv–xxxiii.

Whitwell JL, Duffy JR, Strand EA, Machulda MM, Senjem ML, Gunter JL, Josephs KA. Neuroimaging comparison of primary progressive apraxia of speech and progressive supranuclear palsy. European Journal of Neurology. 2012; 20:629–637. [PubMed: 23078273]

Wilson SM, Henry ML, Besbris M, Ogar JM, Dronkers NF, Jarrold W, Gorno-Tempini ML. Connected speech production in three variants of primary progressive aphasia. Brain. 2010; 133:2069–2088. [PubMed: 20542982]

Yorkston KM. Treatment efficacy: dysarthria. Journal of Speech, Language, and Hearing Research. 1996; 39:S46–S57.

Yorkston, KM.; Miller, R.; Strand, EA.; Britton, D. Management of speech and Swallowing in Degenerative Neurologic Disease. 3rd Ed.. Pro-Ed; Tucson, AZ: 2012.

Youmans G, Youmans SR, Hancock AB. Script training for adults with apraxia of speech. American Journal of Speech Language Pathology. 2011; 20:23–37. [PubMed: 20739633]

Table 1

Types, loci, and distinguishing characteristics of motor speech disorders.

MOTOR SPEECH DISORDER TYPE	Locus	Chief distinguishing auditory perceptual characteristics	Distinguishing pathophysiology
Dysarthrias	Central or peripheral nervous system	Dependent on type	Dependent on type
Flaccid*	Lower motor neuron	Breathiness; hoarseness; short phrases; audible inspiration; hypernasality; audible nasal emisison; imprecise articulation	Weakness
Spastic	Upper motor neuron (usually bilateral)	Strained-harsh voice quality; monopitch & monoloudness; slow rate	Spasticity
Ataxic	Cerebellar control circuit	Irregular articulatory breakdowns; telescoping of syllables; distorted vowels; excess & equal stress; inappropriate variation in pitch, loudness & duration	Incoordination
Hypokinetic	Basal ganglia control circuit	Reduced loudness; monopitch & monoloudness; rapid or accelerated rate; short rushes of speech; reduced stress; repeated sounds; inappropriate silences	Rigidity; bradykinesia; reduced range of motion; movement scaling problems
Hyperkinetic	Basal ganglia control circuit	Variable, dependent on locus in speech mechanism of fast-to-slow and rhythmic-to-unpredictable movements	Involuntary movements (e.g., chorea, myoclonus, tics, dystonia, tremor)
Unilateral UMN	UMN, unilateral	Harshness; hoarseness; imprecise articulation; irregular aticulatory breakdowns; slow rate	UMN weakness; ? spasticity; ? incoordination
Apraxia of speech	Left hemisphere (predominantly frontal lobe)	Distorted sound substitutions & additions; syllable segregation; increased interword intervals; slow rate; articulatory groping; reduced accuracy with increased utterance length & complexity	Disturbed planning/programming of speech movements

*Specific auditory perceptual characteristics that characterize flaccid dysarthrias vary as a function of the specific lower motor neurons that are involved (e.g., specific cranial and spinal nerves).

Table 2

Motor speech disorders associated with PPA variants and related neurodegenerative diseases.

	Dysarthrias						Apraxia of Speech	Typical Underlying Pathology
	Flaccid	**Spastic**	**Ataxic**	**Hypokinetic**	**Hyperkinetic**	**Unilateral UMN**		
PPA								
nfvPPA	-, +/−	-, +/−	-	-, +/−	-	-, +/−	+	Tau (PSP or CBD)
lvPPA	-	-	-	-, +/−	-	-	-, +/−	Alzheimer's disease
svPPA	-	-	-	-	-	-	-, +/−	TDP-43
PPAOS	-, +/−	+/−,+	-, +/−	+/−	-	+/−	++	Tau (PSP or CBD)
Related Diseases*								
PSP	-	+/−, +	+/−	+	-, +/−	-	-, +/−	Tau (PSP)
CBS	-	+/−	+/−	+/−	+/−	-, +/−	+/−	Tau (CBD)
MND/ALS	+	+	-	-	-	-	-, +/−	TDP-43

PPA: primary progressive aphasia

nfvPPA: agrammatic/nonfluent PPA

CBS: corticobasal syndrome

CBD: corticobasal degeneration

lvPPA: logopenic PPA

svPPA: semantic variant PPA

PSPS: progressive supranuclear palsy syndrome

PSP: Progressive supranuclear palsy

MND/ALS: motor neuron disease/amyotrophic lateral sclerosis

++ = always present

+ = frequently present (majority)

+/− = may be present but not in a majority of individuals

- = never or rarely present

-,+/− or +/−,+ = frequency varies within the given range

[i] Some studies of progressive nonfluent aphasia probably include cases that could be classified as lvPPA. This likely leads to an underestimate of the frequency of dysarthria and AOS in the nfvPPA variant.

[ii] Clark et al. (2005) reported that 78% of 26 “nonfluent” cases were “dysarthric.” We did not include this in our tabulations because we suspect the figure included cases with AOS who may or may not have been dysarthric.

[iii] There may be duplicate representation of subjects among studies from the same research groups that could distort these composite estimates of dysarthria and AOS prevalence.

[iv] The greater prevalence of parkinsonian or extrapyramidal features (e.g., bradykinesia, rigidity) in people with nfvPPA/progressive nonfluent aphasia than lvPPA may be helpful diagnostically when they cannot be distinguished on the basis of speech and language features alone (Graff-Radford et al., 2012; Kremen, Mendez, Tsai, & Teng, 2011; Roh et al., 2010).

[*] Motor speech disorder frequency is for the typical presentation of these disorders, not when PPA or AOS are early or predominant signs.