How Evidence-Based Practice (E³BP) Informs Speech-Language Pathology for Primary Progressive Aphasia

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Abstract
Amelioration of communication impairment in primary progressive aphasia (PPA) is an area of clinical importance and current research. Speech-language pathologists (SLPs) have a range of skills and interventions to support communication in PPA; however, underrecognition of their role and low referral rates is an ongoing concern. The E³BP conceptualization of evidence-based practice comprises 3 components: research-based evidence, practice-based evidence, and informed patient preferences. Here, we will describe how evidence for managing the communication difficulties experienced by individuals with PPA exists at all 3 levels of the (E³BP) model, highlighting how this allows SLP interventions to be both evidence based and patient centered. We encourage health professionals to value and utilize the wide range of services that SLPs can offer when working with individuals with PPA, to educate, remediate everyday linguistic skills, increase daily participation, and maximize overall quality of life.

Keywords
primary progressive aphasia, evidence-based practice, evidence-based medicine, speech and language pathology, communication

Introduction
Dementias that are characterized predominantly by deterioration in communication skills are known collectively as “language-led dementias” or “primary progressive aphasia” (PPA). Primary progressive aphasia was first defined as “slowly progressive aphasia” by Mesulam,1(p592) and diagnosis requires insidious language decline.2 Unlike amnestic forms of dementia, however, individuals with PPA in the initial to mid stages typically retain their nonlinguistic cognitive skills (including episodic memory) and often demonstrate acute awareness of their communication difficulties.3,4

Primary progressive aphasia is not a disease per se, it is a range of heterogeneous clinical syndromes that do not have one underlying cause or pathology. As detailed by the current international diagnostic criteria,2 there are 3 main and widely accepted clinical presentations: semantic variant, nonfluent/agrammatic variant, and logopenic variant.2 All 3 variants typically have word-finding difficulties and impaired language production but differ in the nature of other linguistic impairments such as impaired language and object comprehension in semantic variant PPA and motor speech coordination impairments in nonfluent/agrammatic variant PPA.5 In the later stages of the condition and as the neuropathology progresses, additional cognitive, behavioral, and motor symptoms emerge.5

Primary progressive aphasia was previously described as the language presentation of frontotemporal dementia (FTD) with 2 clinical variants: a semantic and a nonfluent variant, until Gorno-Tempini and colleagues introduced the logopenic variant, and the need for consensus criteria for PPA diagnosis was recognized.2,6,7 Since the development of the consensus criteria, however, we have come to understand that around 1 in 5 cases will remain difficult to categorize.4 The changing clinical and pathological characterizations of PPA over the years have made it challenging for clinicians to keep current with information about PPA.6,9 Notably, while some variants of PPA (semantic and nonfluent) typically arise from frontotemporal lobar degeneration (FTLD; note 1), others (logopenic

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variant) have Alzheimer’s pathology and are considered as atypically presenting Alzheimer’s disease. There are no studies establishing the prevalence of PPA per se, and calculations are muddied by the fact that most studies of FTD prevalence do not include the logopenic variant PPA (but do include motor and behavioral FTLD syndromes). However, Marshall and colleagues have suggested that PPA prevalence may be roughly in the order of 3 per 100,000. The fact that PPA has only been recognized since the early 1980s, and knowledge of the PPA variants, their clinical presentations and pathologies is still expanding, means that research into the role of speech-language pathologists (SLPs) and communication intervention for people with PPA is relatively in its infancy compared with stroke aphasia.

On a day-to-day basis, individuals with PPA will experience difficulties with communication in a variety of different contexts. These might range from social situations to service transactions, from communicating clear financial and legal decisions to making basic needs or preferences understood. Often individuals can maintain independence in these areas for longer with adequate and individualized support from health professionals such as SLPs. Even SLPs who have not had the benefit of specific training in PPA possess extensive knowledge of the assessment and management of communication, cognitive rehabilitation, activities of daily living, and designing person-centered interventions, and this makes them uniquely positioned to enhance the communication, coping, and quality of life of persons with PPA. Nevertheless, PPA referral rates to SLP services remain low and the role of SLPs working with communication in dementia syndromes is underrecognized. Speech-language pathologists also provide vital support for swallowing difficulties in dementia syndromes (see the studies for further discussion of this issue); however, this article focuses on communication support.

Individuals with PPA may present to a wide range of care providers such as medical specialists, general practitioners, nurses and care workers, as well as SLPs. The aim of this article is to increase awareness among these health professionals that effective communication interventions are available to people with PPA and their families because SLPs are trained to develop intervention plans by triangulating 3 sources of evidence: research evidence, practice-based evidence, and the preferences of a fully informed patient. This “multiple sources of evidence” approach is consistent with emerging emphases on patient-led care, quality of life, and patient-reported outcome measures in health-care systems globally. Comprehensive reviews of treatment options in PPA are available elsewhere, and interested readers are encouraged to consult these for a summary of the latest research evidence.

**Provision of Communication Intervention for PPA**

Historically, rehabilitative-style interventions have often not been offered to individuals with PPA, or indeed any dementia syndromes. This has, first, been related to “the traditional therapeutic nihilism that frequently accompanies diagnosis of progressive disorders” that may be adopted when managing individuals with PPA. Second, referring parties continue to have low awareness of the value of SLP in positive management of the communication needs of people with PPA. In fact, the full extent of underrecognition and underreferral is unknown. Clearly, education for all health professionals on the role of the SLP in PPA management is of ongoing importance to ensure the best outcomes for these individuals. Indeed, some barriers to services for people with PPA have also come from within the SLP profession itself. The first dementia position paper from the Royal College of Speech Language Therapists in the United Kingdom, for example, was as recent as 2005. It focused on memory difficulties in dementia and did not address PPA at all. Subsequently, an Australian study in 2009 found that SLP respondents reported lack of confidence in dealing with PPA and a US study in 2014 revealed that 43% of SLPs reported not being familiar with PPA at all.

Nowadays, a pressing amount of evidence exists to demonstrate that provision of communication intervention for PPA is valuable and critical for several reasons. First, PPA can be a younger onset dementia: individuals with PPA may be in their 40s or 50s, busy raising families and/or working. In fact, research has shown that FTDS, including PPA, are associated with a substantial social and economic cost to individuals and their families. Second, wide variation exists in survival and prognosis. Average survival has been reported as 10.6 years for people with semantic variant or 8 years for individuals with nonfluent variant following the onset of symptoms. Language decline may be sufficiently slow in some cases that individuals live with relatively focal language deficits for over 10 years. In other cases, however, the progression from purely linguistic to more generalized impairments can be markedly faster (note 2; see case described by Mahendra) For example, compare S1 and S5 in Croot and colleagues’ study, both of whom had initially similar presentations of nonfluent variant PPA: one deteriorated quickly after diagnosis and over a matter of months, while the other showed minimal change in mood and nonlanguage cognitive abilities over a period of years, resulting in considerable differences in their candidacy for interventions. Third, many individuals with PPA have a heightened awareness of their communication impairments as their insight is typically intact, which can be associated with clinically significant distress and depression. In summary, whether the language decline is rapidly progressive or painfully slow, each of these factors strongly warrants targeting lost linguistic skills and supporting important communicative interactions in people with PPA, enabling them to maximize their communication, autonomy, and quality of life for longer.
families also benefits relationships, coping strategies, mental health and quality of life.18,19

Moving forward, the words of Murray in 1998: “Professionals such as neurologists, neuropsychologists, and speech-language pathologists are encouraged to advocate for the provision of services to patients with PPA,33(p669) through to those of Cartwright more recently in 2015: “It is important that clinicians have the skills and resources available to instill hope and foster proactive coping strategies as a key component of therapeutic management of PPA.”17(p188) continue to ring true. Although the translation of this message into clinical services has been slow at times, this message is nonetheless being disseminated more and more, particularly in expert opinion pieces.36,42,45,46 If we are mindful that people with PPA have specific needs that may not be fulfilled by services for amnestic dementias, the promotion of SLP interventions for individuals with dementia by professional associations is also promising. Practice statements now highlight that SLP interventions for individuals with dementia should be based on communicative strengths and weaknesses48 and “promote and maintain [the] functional and social independence of people with dementia”.49(p7) Last, it is positive to see the popularity of learning opportunities and professional development workshops on PPA. Both the last 2 International Conferences on Frontotemporal Dementias have incorporated workshops to provide education on PPA to a wide range of clinical service providers, including SLPs, together with publicly available education resources.50

Sources of Evidence for Communication Intervention in PPA

The E3BP Approach

Evidence-based medicine was first defined by Sackett and colleagues26 and further elaborated as evidence-based practice for SLPs.26,52 The latter is often described using the abbreviation E3BP,26 making it explicit that there are 3 sources of evidence that are equally relevant in clinical decision-making. We define the 3 types of evidence here largely following Dollaghan and LoF.26,52 Research evidence refers to the best available evidence from systematic research with high internal validity that applies to a given clinical question.26 Practice-based evidence has also been previously described as “clinical expertise”51(p71) and “evidence internal to clinical practice.”26(p2) This refers to evidence which is collected in a clinical setting and which can have greater real-life application than is the case for some tightly controlled studies which generate research evidence.52 Preferences of a fully informed patient refers to how clinicians develop a shared understanding of the “beliefs, preferences, hopes and fears”26(p2) of individuals in their care so that these can be used to select interventions and set goals and therapeutic targets that the individuals will have confidence in and comply with. Dollaghan26 discusses how best to obtain this information when working with individuals with communication difficulties, for example, via interview, proxy interview, or evidence triangulation.

Research-Based Evidence for SLP Services in PPA

Is there research evidence that SLPs can provide efficacious intervention to individuals with PPA? There are now numerous studies demonstrating the impact of interventions for PPA which focus directly on practicing and maintaining communication skills. These include systematic reviews (see studies29-31 for an overview) and an increasing number of single case-experimental design studies, both considered to provide high-quality evidence by the Oxford Centre for Evidence-Based Medicine,53,54 as well as a growing body of case-controlled group studies. The majority of reports investigate lexical retrieval treatment, which is also described as word relearning.30 These interventions train individuals with PPA to better retrieve a core vocabulary of personally relevant words. Many individuals benefit greatly from such training, which is evidenced by better retrieval of trained words than untrained words. Additionally, many of the individuals who participate in this type of intervention find it rewarding and positive.55 While a few studies have demonstrated generalization to untrained items or contexts, there is no clear understanding yet of the mechanisms or factors which might promote generalization.56-58 Script training, which promotes rote learning of personally relevant statements and monologues,59 and discourse training, which supports comprehension60 and/or production of sentences and stories,61,62 have also been shown to improve communication abilities in early to mid-disease for individuals with PPA. Although such interventions cannot prevent inevitable language deterioration, these studies provide research evidence that learning of practiced items and/or new strategies is indeed possible in PPA.

A number of other studies have found positive outcomes when supporting individuals to continue to engage in daily activities. Examples include facilitating comprehension and recall of, and discussion about, TV shows,60 learning to write notes with accompanying photos in a smartphone to aid recall of shopping lists or to provide recounts of recent activities,63,64 and using additional written and visual cues to aid in following recipes.65 Improvements following such interventions are not only constrained to research settings but have resulted in spontaneous use of trained skills in novel everyday settings.64

In addition to the options available on an individual basis, SLPs offer education, support, and/or conversation/communication groups to individuals with PPA and their families. Qualitative comments and survey data indicate that individuals with PPA and their families welcome education about PPA.55,66 They also appreciate both direct psychosocial support and, indirectly, the opportunity to reduce isolation and meet others with the diagnosis66-68 that such groups provide. They also value the ability to practice communication skills in the naturalistic group setting and/or with new communication partners.63 Family and communication partners can learn to better facilitate successful conversation.55,68,69 Not only do
participants report positively on many aspects of group intervention but there is additional evidence that these activities improve quality of life and/or confidence. As they typically include multiple components concurrently, it remains an open question which of the components of these group interventions contribute most to their positive outcomes.

Interventions can successfully be delivered via different modalities and platforms: face-to-face in the clinic, remotely using telephone or online platforms, by a trained primary communication partner at home, or by the individual with PPA working on home treatment tasks.

**Practice-Based Evidence**

Practice-based evidence can include expert opinions, in-practice data collection, case reports, or even family reports on the successes and challenges of interventions at home. Speech-language pathologist expert opinions and commentaries which advocate for particular interventions reflect the authors’ perceptions that those interventions have been effective in their clinical practice. For example, the majority of clinicians working with PPA in the United Kingdom reported that they predominantly use interventions which aim to increase participation in everyday goals and activities. A number of other expert opinion papers or textbooks also advocate for this approach. Interventions that have been suggested to be supported by practice-based evidence include teaching conversation repair strategies and/or circumlocution (to help individuals communicate their message when they are unable to find the exact words they want), communication partner training (where partners are taught how to communicate in ways which facilitate successful exchanges and reduce frustration), multimodal communication (where any modality such as writing, drawing, gesturing, and/or speaking can be used to convey a message), and the use of communication aids (selecting symbols and/or pictures to convey a message).

Case studies of individuals with PPA who have demonstrated personally and clinically meaningful gains following intervention also provide practice-based evidence. For example, Murray describes a woman with PPA and her husband who experienced various communication benefits from multimodal communication, communication partner training, and use of an electronic communication aid over time, as well as social benefits from participation in an aphasia support group.

Lastly, the stories of those with PPA and their families regarding the effectiveness and usefulness of certain interventions is not to be overlooked as practice-based evidence. One lay author describes the significant impact a digital text to speech communication device had for her husband: “It gave Boyd his quality of life back again and he had confidence now to talk to people through the machine. It was definitely the right time for him to have it while he still had the cognitive skills to use it. He settled down again from being frustrated and agitated and was happier in himself for a while.” Reports such as these give SLPs knowledge about which interventions can benefit which individuals and can assist in determining whether a subsequent patient is similar enough to be likely to benefit from the same approach.

**Preferences of Informed Patients**

Integrating the preferences of informed patients is the third source of evidence in E3BP. Shared decision-making to align intervention approaches, goals, and specific items for therapy with patient preferences on a case-by-case basis is also consistent with a “patient-centered approach.” Many individual factors may predispose a person with PPA to enjoy, benefit from, or engage with a particular intervention, including their degree of motivation, family support, choice of goals, current mood, cognitive resources, potential for learning and change, financial resources, and logistical issues such as transportation. Taking these factors and preferences into account is likely to improve patients’ motivation and engagement with services, as well as their confidence and quality of life.

Once a patient has trialed an intervention, SLPs can consider questions such as: What did the patient think of the intervention? Is it something they feel they would commit to over a longer term? Do the specific goals or items being targeted need to be adjusted? Indeed, given that individual responses to interventions cannot be guaranteed in advance, some PPA researchers are recommending short-term trials of treatment as indicators of who might be motivated and helped by treatment. In addition, examining the preferences of patients should include consideration of their linguistic and sociocultural background so that interventions and therapy targets can be aligned and delivered in an affirmative and culturally congruent manner.

**Triangulating Evidence Sources**

E3BP integrates all three forms of evidence during clinical decision-making. Once informed about the options, every individual will have different priorities for their communication and these need to be balanced against available research evidence and practice-based evidence. For example, research evidence tells us that word relearning interventions can be effective for core vocabulary as well as increasing confidence and participation. We have also learned that some research participants are highly motivated to improve their speech and/or language impairment. For example, in our own research, a 64-year-old participant with logopenic variant PPA said, “You have to do things that are going to help you talk. . . . Finding the words is the best thing you can do.” Combining research evidence and patient preferences suggests that lexical retrieval treatment would be an excellent choice to support word-finding by the first individual, but combining practice-based evidence and patient preferences suggests it would be unsuitable for some others, despite the research evidence. Similarly, research evidence tells us that introducing alternative modalities of communication earlier in the progression of the disease can relieve frustration and increase independence for some individuals. In contrast, others may find that being
introduced to compensatory-style interventions uncomfortably highlights the future reality that their language will decline.\textsuperscript{36} Taking both the research evidence and the patient preferences into account, the SLP might decide not to introduce this type of intervention for the latter individuals.

Ultimately, the SLP needs to make a final decision about how to proceed, incorporating individual differences and all intervention options. Given that SLPs are trained to ensure that options are explained in an accessible and communication-friendly manner, individuals with communication impairments should be given every opportunity to engage in this decision-making process. The examples provided show how E\textsuperscript{3}BP is a useful framework for integrating research and practice-based evidence, as well as the preferences of a fully formed patient, by weighting different types of evidence on a person-by-person basis.\textsuperscript{81}

**Conclusion**

Individuals with PPA experience devastating loss of communication skills, often while they are fully aware and are still in a very active phase of their lives. The effect of communication changes on quality of life, engagement in everyday activities, as well as families and relationships is extensive. Those of us working clinically with PPA need more research on the disease trajectory, treatments (effectiveness, candidacy, mechanisms underlying treatment gains, and generalization), compensation for communication difficulties (such as the forthcoming work being conducted in conversation partner training),\textsuperscript{82} and a better understanding of perspective of individuals with PPA and what factors can promote well-being.\textsuperscript{42}

Speech-language pathologists can, however, already do a considerable amount to maximize day-to-day communication, participation, relationships, and quality of life in PPA. The 3 components of evidence-based practice—research-based evidence, practice-based evidence, and the preferences of informed patients—allow for a robust and broad range of evidence to be considered when supporting individuals with PPA with their communication, warranting early referrals to SLP services. It is crucial that all health professionals involved in the management of individuals with PPA at all levels of care, from diagnostic specialists to late-life residential care providers, are aware of the value of communication support and intervention for these individuals and their families.

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**Notes**

1. Frontotemporal lobar degeneration (FTLD) is a spectrum of neuropathologies associated with various clinical presentations including semantic variant primary progressive aphasia (PPA), nonfluent variant PPA, behavioral variant frontotemporal dementia, or motor syndromes including corticobasal syndrome and progressive supranuclear palsy.\textsuperscript{18} Its progression is generally faster than that of Alzheimer’s disease.\textsuperscript{43} Only the PPA variants are discussed in this paper (see the studies\textsuperscript{9,84-86} for further discussion of clinical presentations, prognosis, and management of FTLD).

2. It is worth noting that the ability to assess whether solely linguistic versus more generalized cognitive functions are impaired is marred by the fact that many of the available and widely used assessments (such as the Mini Mental State Examination\textsuperscript{87} or Addenbrooke’s Cognitive Examination III\textsuperscript{88}) rely heavily on language skills including comprehension, semantics, and expression. In such tests, the occurrence of word-finding difficulties, for example, can result in a reduced score on a memory or orientation subsection.

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