
Managing Primary Progressive Aphasia (PPA)

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Editor's Note: *In layperson's terms, primary progressive aphasia (PPA) is a slowly worsening aphasia not due to stroke, trauma, tumor or infection. Although PPA is considered relatively rare, we regularly receive inquiries about the condition. The National Aphasia Association (NAA) is very grateful to Joseph Duffy for the following submission.*

Primary progressive aphasia (PPA) can leave affected persons and their loved ones with many questions and a sense of helplessness about the decline in the ability to communicate verbally and to read and write. These feelings may have been present for some time, but once the diagnosis of PPA is made, the questions tend to shift from "What is this problem?" to "What now? What can be done about this?"

By definition, PPA is progressive, but it is natural to ask what can be done to improve or maintain the ability to understand and express language. These questions cannot yet be answered with certainty. Although hundreds of cases have now been reported in the literature and thousands of people are probably affected, PPA is considered a rare condition. In addition, it may have more than a single underlying cause and there are few reports about the effectiveness of treatment for it. Nonetheless, it appears that some steps can be taken to help manage the communication problems.

The following overview offers some basic guidelines expressed in general terms. Because the severity and nature of the communication difficulties vary greatly, any treatment must be tailored to the particular individual.

The Medical Perspective

Most approaches to treating aphasia can be categorized as either medical or behavioral. From the medical perspective, there are currently no drugs or other interventions specifically designed for PPA. This partly reflects our limited understanding of what causes PPA and the likelihood that it has more than a single cause. Neurologists sometimes prescribe drugs that are used for people with Alzheimer's disease under the assumption

aphasia

that PPA and Alzheimer's disease may share a common cause. This assumption is unlikely to be true in the majority of cases. Currently, there are only anecdotal reports that the same drugs that target Alzheimer's disease are helpful in relieving the signs and symptoms of PPA.

The Behavioral Perspective

The primary approaches to managing PPA at this time are behavioral. That is, there are things that the person with PPA can do that may lessen the impact of the disease. Behavioral approaches emphasize practice, drill, and counseling to (1) enhance the ability to communicate, or (2) compensate for the inability to communicate in conventional ways.

Some behavioral approaches for PPA are directed at improving or maintaining (in the short term) impaired language abilities. The decision to pursue this type of therapy should take into account the following considerations. First, the person with PPA must still have some capacity for insight, motivation and learning. Without them the possibility of meaningful improvement is greatly reduced. Also, the individual's significant others must be motivated and involved as well. They play an important role in working on practice activities beyond formal therapy sessions and in providing cues for using effective communication strategies. Finally, everyone involved must understand that therapy will not eliminate difficulties with communication. Even if there is improvement in communication ability, it will not reverse the progression of the disease.

The skills targeted for therapy are generally based on three factors: those abilities that are declining, those that may be relatively preserved, and those that are most important to the affected person. In all instances, therapy requires work that might be called "focused exercise of the brain's language system."

At this time a small number of reports - all of which are based on only one person or a few carefully selected people with PPA - have documented improvements in abilities targeted by therapy. Skills that have improved include comprehension of spoken instructions and questions, production of sentences, retrieval of words, and number reading. It is premature to conclude that such treatments are likely to be effective for many people with PPA. However, these reports do suggest that for some affected individuals, and for some deficits, therapy may be beneficial. Whether these benefits continue beyond the period of formal therapy is not known.

Compensatory Strategies for both Patients and Families

Other behavioral approaches emphasize compensatory strategies that can improve communication, although not necessarily in conventional ways. Compensatory strategies can be "patient-oriented" or "other-oriented," or a combination of both. Patient-oriented strategies reflect things that the person with PPA can do to enhance communication, such as establishing the topic at the outset of a conversation, using gestures, and using pictures, writing or



aphasia

drawing. "Other-oriented" strategies include paying full attention to the affected person, giving feedback about the need for clarification, providing more time for communication, confirming information, keeping statements relatively brief, and supplementing speech with gestures. Joint efforts on the part of both the person with PPA and others include speaking in environments that are conducive to effective communication (such as face-to-face conversations with minimal noise and other distractions). People with PPA and their significant others often benefit from following the rule that communicating requires everyone's full attention. The notion that people with PPA require all the fuel in their "language tank" when communicating is a useful analogy and one to keep in mind when engaged in speaking, listening, reading or writing activities.

It can be very helpful to consult with a speech-language pathologist (SLP) to identify important communication needs, learn how and when compensatory strategies can best be used, and practice their use. The SLP can help to identify specific strategies and investigate whether augmentative strategies (for example, gesture, pantomime, and drawing) may supplement or sometimes replace verbal communication. Such strategies have been reported as helpful in some people with PPA.

Electronic/computer devices may be able to supplement or replace speech in some people with PPA. Because they require a person to use conventional language or other symbols, they may not be helpful for those whose language skills are already severely impaired. Some people with PPA also have an apraxia of speech (AOS, a problem with the programming of movements for speech rather than a language problem). They may have speech that is far more impaired than their language comprehension or ability to read and write. As long as their ability to control movements of body parts needed to use the devices is relatively intact, those with AOS and relatively mild PPA may be good candidates for electronic/computer alternatives to speech. Generally, the development, practice and learning of augmentative or alternative means of communication should occur well before there is an actual need to use them, so they are readily available and more easily used when and if the need emerges.

Finally, there can be little doubt that simply learning about PPA is beneficial, both psychologically and practically. As discussed above, SLPs who have experience working with people with aphasia and degenerative neurological diseases can address questions about aphasia in general, and PPA in particular, and can help plan for future communication needs. Free or low-cost evaluations and therapy may be available from university speech-language clinics. Some PPA patients and/or spouses benefit from joining an Aphasia Community Group or Stroke Support Group that has others with aphasia in it. This is true even if the others do not have aphasia that is progressive. The National Aphasia Association (NAA) provides information about PPA and listings of support groups (www.aphasia.org). The American Heart Association resources can make referrals to stroke support groups (www.americanheart.org).

Viewed in the most positive sense, the diagnosis of PPA does not mean the end of communication. It can be the first step to identifying ways to maintain communication abilities for as long as possible. ♦