Primary progressive aphasia

What is Primary Progressive Aphasia (PPA)?
Primary Progressive Aphasia (PPA) is a neurological syndrome in which language capabilities become slowly and progressively impaired. Unlike other forms of aphasia that result from stroke or brain injury, PPA is caused by neurodegenerative diseases, such as Alzheimer’s Disease or Frontotemporal Lobar Degeneration. PPA results from deterioration of brain tissue important for speech and language. Although the first symptoms are problems with speech and language, other problems associated with the underlying disease, such as memory loss, often occur later.

PPA commonly begins as a subtle disorder of language, progressing to a nearly total inability to speak, in its most severe stage. The type or pattern of the language deficit may differ from patient to patient. The initial language disturbance may be fluent aphasia (i.e., the person may have normal or even increased rate of word production) or non-fluent aphasia (speech becomes effortful and the person produces fewer words). A less common variety begins with impaired word-finding and progressive deterioration of naming and comprehension, with relatively preserved articulation.

As with aphasia that results from stroke or brain trauma, the manifestations of PPA depend on what parts of the left hemisphere are relatively more damaged at any given point in the illness. The person may or may not have difficulty understanding speech. Eventually, almost all patients become mute and unable to understand spoken or written language, even if their behavior seems otherwise normal.

Signs and symptoms of other clinical syndromes are not found through tests used to determine the presence of other conditions. PPA is not Alzheimer’s disease. Most people with PPA maintain ability to take care of themselves, pursue hobbies, and, in some instances, remain employed.

Is there any treatment or assistance for people with PPA?
People with primary progressive aphasia are fighting against a condition in which they will continue to lose their ability to speak, read, write, and/or understand what they hear. Usually people with aphasia that results from stroke or head injury will experience improvement over time, often aided by speech therapy. This is not the case for people with primary progressive aphasia. However, individuals with PPA may benefit during the course of their illness by acquiring new communication strategies from speech-language pathologists. Some families have also learned new strategies through participation in Aphasia Community Groups.

Many people with aphasia find it helpful to carry identification cards and other materials that can help explain the person’s condition to others. ID cards are available from the National Aphasia Association website. Some communication-assistive devices may also be helpful. Non-verbal techniques for communicating, such as gesturing and pointing to pictures, may help people with PPA express themselves.

Managing primary progressive aphasia
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 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 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.

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