

Department of Clinical Neuropsychology

Understanding Primary Progressive Aphasia



Primary progressive aphasia (PPA) is a form of worsening (“progressive”) language dysfunction. “Aphasia” is a general term applied to language disorders that begin in adults because of brain injury or disease.

Symptoms

Early symptoms of PPA often include difficulty coming up with words, correctly pronouncing words, or understanding words, despite normal thinking ability in other areas (like memory and spatial abilities). At first, language is the only aspect of thinking affected. Later, other mental abilities—such as memory and reasoning—also become impaired. The exact symptoms a person has depend on what kind of PPA they have.

Types

Logopenic (PPA-L)

PPA-L is characterized by prominent **problems with word-finding**. The word *logopenic* means “lack of words.” Individuals with PPA-L may “talk around” the word they cannot find or may substitute another word. They may also resort to filler statements such as “you know” or “that thing.” Frequent pauses in speech, mispronunciations, and spelling errors are also common. Over time, a person with PPA-L is likely to have difficulty understanding long or complex sentences.

Agrammatic/Nonfluent (PPA-G)

PPA-G is characterized by prominent **problems with speech production**, resulting in short sentences and frequent hesitations. People with PPA-G may mispronounce words or misuse them. Sometimes they say words in the wrong order. Frequent pauses and mispronunciations reduce the overall smoothness, or *fluency*, of the person’s speech. Someone with PPA-G may also begin to omit function words (“and” and “but”) and pronouns (“she” or “he”), resulting in “agrammatic” speech. Understanding single words is usually normal, but over time, a person with PPA-G may struggle to understand long or complex sentences.

Semantic (PPA-S)

PPA-S is characterized by prominent **problems with language comprehension and naming objects**. A person with PPA-S can still speak fluently but will lose their ability to understand simple words such as “dog” or “boat.” Nouns (names of things) are more affected than other

words and may begin to disappear from speech. In most people, PPA-S affects the left side of the brain. However, in rare cases, the disease develops on the right side of the brain. PPA-S on the right side leads to impaired visual recognition of familiar things, usually faces. Individuals with PPA-S may also have behavioral problems, such as irritability, loss of empathy, or poor judgment.

Causes

PPA is caused by cell death or *neurodegeneration* in the parts of the brain that control speech production and language abilities. Language is usually managed by the left side of the brain. So, PPA almost always involves degeneration of the left side.

In the most cases, PPA is “sporadic,” which means it can happen to anyone. It can even happen to people with no family history of dementia or other neurological diseases. In rare cases, PPA is caused by hereditary gene mutations. Both sporadic and hereditary PPA cause a buildup of protein in some parts of the brain, which leads to degeneration.

PPA-L is usually caused by the same process as Alzheimer’s disease, so it is known as an “atypical” form of Alzheimer’s disease. Like typical Alzheimer’s disease, people with PPA-L often eventually develop memory loss and spatial disorientation.

PPA-G and PPA-S are closely related to a form of dementia known as frontotemporal lobar degeneration or FTLT. People with FTLT show changes in motor control and social behavior. Some of these symptoms may also be seen in patients with PPA-G and PPA-S.

Risk Factors

- **Age:** PPA is most common in those over age 65.
- **History of dyslexia:** Rates of dyslexia are higher in PPA patients and their first-degree relatives.
- **Genes:** Certain genes associated with increased risk for FTLT and dyslexia are more common in individuals with PPA.
- **Brain injury:** A history of injury to the left side of the brain, even if it occurred many years before the onset of PPA symptoms, has been linked to increased risk for PPA in a small number of cases.

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Common Diagnostic Tests

The diagnosis of PPA usually begins with an appointment to see a neurologist. The neurologist asks questions to get details on current symptoms and past functioning. Additional testing, such as neuropsychological testing and MRI of the brain, is often done too.

Neuropsychological testing relies on pencil and paper and computerized tests to look at thinking abilities, including how language function is impaired. When neuropsychologists interpret the test results, they look for patterns of language dysfunction that suggest which parts of the brain are most likely affected. Some of these changes are not visible on MRI.

An MRI can show structural changes in the brain, such as areas of shrinkage, which can further support a particular diagnosis. Occasionally, a special scan known as a PET scan can provide information about how the brain is functioning.

Although brain scans help the neurologist diagnose PPA, it is ultimately the pattern of language dysfunction that is used to determine the type of PPA. Other disorders, including stroke and psychiatric illnesses, can also cause changes in language function that are not considered PPA.

Treatment

There are no medicines yet to treat PPA. Because PPA-L is related to Alzheimer's disease, medicines used to treat Alzheimer's disease are sometimes prescribed. However, none have been shown to improve the symptoms of PPA. When necessary, other medicines may be prescribed to manage behavioral symptoms such as depression or agitation.

Speech therapy is frequently recommended to improve a person's ability to communicate. It is particularly helpful in the early stages of PPA. Treatment may involve teaching ways to compensate for the symptoms, such as techniques to enhance word-finding ability or speech production, or to use augmentative or alternative communication devices. Family members are encouraged to take an active role in treatments.

Suggestions for Patients

- Carry a card with written identification and a brief description of your language impairment to help strangers identify and assist you.
- Ask for written information, especially if it is new or unfamiliar (for example, when listening to your doctor's advice).
- Continue to do activities you enjoy, attend cultural events, read, exercise, and socialize as much as possible.

- Create an organized environment. Arrange items in a particular place and always put them back there. This minimizes stress and may be very helpful if you begin to have changes in your memory.
- Adhere to a routine as much as possible. Consistent habits help you remember to take your medication every day, particularly if you have memory decline.

Suggestions for Caregivers

- Avoid pointing out mistakes.
- Avoid speaking for the person. Develop nonverbal cues to indicate when help is desired.
- Ask closed questions, for example, *"Do you want tea or coffee?"* instead of open questions like *"What do you want to drink?"*
- Have the speaker describe a word they cannot find by asking closed questions about its function (*"Is it for hammering?"*), or appearance (*"Is it blue?"*).
- Listen for key information (who, what, where, when, why, and how).
- Eliminate distracting background noise whenever possible.
- Use written options and allow the person with PPA to point to the choice they want.
- Develop a system of gestures to communicate everyday needs and functions *before* these gestures are actually needed.
- Use body language (eye contact, facial expressions, pat on the back, hugs) to communicate support.

Resources

Additional resources for patients and caregivers, such as educational materials, support groups, and more, can be found below.

The National Aphasia Association

www.aphasia.org

The Association for Frontotemporal Degeneration

www.theaftd.org

The Alzheimer's Association

www.alz.org

UCSF Memory and Aging Center

memory.ucsf.edu/dementia/primary-progressive-aphasia

Mesulam Center for Cognitive Neurology and Alzheimer's Disease

www.brain.northwestern.edu/dementia/ppa/symptoms-causes.html