
understanding primary progressive aphasia (PPA)

PRIMARY PROGRESSIVE APHASIA is a rare neurological syndrome in which language capabilities become slowly and progressively impaired. This information sheet was prepared to help explain Primary Progressive Aphasia to the general public.

What is aphasia?

Aphasia is an acquired disorder caused by brain damage which affects a person's ability to communicate. The principal signs of aphasia are impairments in the ability to express oneself when speaking, trouble understanding speech, and difficulty with reading and writing. Aphasia is most often the result of stroke or head injury, but can also occur in other neurological disorders, such as brain tumor or Alzheimer's disease. The effects of aphasia differ from person to person, and can sometimes benefit from speech therapy. Strategies to communicate non-verbally (without words) may also be helpful to the person with aphasia.

What is primary progressive aphasia?

The syndrome of primary progressive aphasia has been defined by Mesulam and colleagues as a progressive disorder of language, with preservation of other mental functions and of activities of daily living, for at least two years. Primary progressive aphasia is not Alzheimer's disease. Most people with primary progressive aphasia maintain ability to take care of themselves, pursue hobbies, and, in some instances, remain employed. The problem is a disorder of language; and signs and symptoms of other clinical syndromes are not found through tests routinely used to determine the presence of other conditions.

Although primary progressive aphasia may take a number of forms, it commonly appears initially as a disorder of speaking (an articulatory problem), progressing to nearly total inability to speak in its most severe stage, while comprehension remains relatively preserved. A less common variety begins with impaired word finding and progressive deterioration of naming and comprehension, with relatively preserved articulation.

However, other neurological disorders exist in which progressive deterioration of language is only one component of a broad, progressive decline of mental functions, including memory, attention, visuospatial skills, reasoning, and the carrying out of complex motor activities. These diseases, such as Alzheimer's disease, Pick's disease, and Creutzfeldt-Jakob disease, should be excluded by appropriate neurologic examinations, when a person experiences progressive language decline.

Is there any assistance for people with primary progressive aphasia?

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,

aphasia

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, although they 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.

For example, aphasia identification cards and other materials available from the the NAA can help explain the person's condition to others. Some communication-assistive devices may also be helpful. Non-verbal techniques for communicating, such as gesturing, pointing to pictures, etc., may help people with primary progressive aphasia express themselves. ♦

Where can I get more information?

Please note: None of the information provided on this fact sheet is intended as medical advice. If you believe you may have primary progressive aphasia, you should consult your physician. For individuals who have access to a medical library or the internet, these papers discuss primary progressive aphasia:

1. Mesulam M-M. Slowly progressive aphasia without dementia. *Annals of Neurology*, 11:592-598, 1982.
2. Weintraub S, Rubin NP, Mesulam M-M. Primary progressive aphasia: longitudinal course, neurological profile, and language features. *Archives of Neurology*, 47:1329-1335, 1990.
3. Kertesz A, Hudson L, MacKenzie IRA, Munoz DG. The pathology and nosology of primary progressive aphasia. *Neurology*, 44:2065-2072, 1994.
4. Kirshner II, Baker M. Syndromes of language dissolution in aging and dementia. *Comprehensive Therapy*, 21:519-523, 1995.
5. Mesulam M-M, Johnson N, Grujic Z, Weintraub S. Apolipoprotein E genotypes in primary progressive aphasia. *Neurology*, 49:51-55, 1997.

Other papers relating to very technical medical issues in PPA are also available. For information on searching through the medical literature, see the NAA's Fact Sheet -Sources of Information about Aphasia Research and Treatment.

PPA Program, Northwestern University. Website <http://www.brain.northwestern.edu/ppa/ppa.html> or contact: PPA Research Coordinator, Northwestern Alzheimer's Disease Center, 675 N St. Clair, 20-100, Chicago, IL 60611, 312-695-2343, 312-695-5747 (FAX), k-simmons@northwestern.edu.

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