

Subtype: Semantic Variant Primary Progressive Aphasia

Semantic PPA is a disorder of language in which patients demonstrate a progressive deterioration of understanding words, especially nouns, and recognizing objects while other cognitive faculties remain remarkably spared. Specifically, patients with semantic PPA retain the ability to produce fluent speech, but without being able to generate the key words within sentences, this speech becomes increasingly difficult to understand. Semantic PPA patients also lose the ability to recognize the meaning of specific words or to spontaneously name familiar, everyday objects.

Unlike other FTD subtypes, semantic PPA generally does not produce changes in behavior or personality until later stages of the disease. Most people with progressive aphasia maintain the ability to care for themselves, keep up outside interests and, in some instances, remain employed for a few years after onset of the disorder.

Key Clinical Features

The hallmark of semantic PPA is difficulty generating or recognizing familiar words. For example, when a patient is shown a picture of a cat, he can neither name it nor can he recognize the word when it is provided. The patient characteristically asks “what is ‘cat’?” when it comes up in conversation or during testing. This happens for rare words first and common nouns for later stages. Verbs and abstract words are surprisingly spared.

Fluent spontaneous speech is retained. Especially in early stages, patients may be able to “talk around” the meaning of a specific word they are otherwise unable to generate. In later stages they can repeat their agenda without listening to other speakers. Word-finding pauses in speech become common, and patients have difficulty naming familiar objects.

Some patients have problems recognizing familiar objects and faces. The presence of this sign can help confirm the diagnosis.

Neuroimaging studies demonstrate loss of brain volume, blood flow or neural activity in the left temporal lobe of semantic PPA patients. The left side of the brain most frequently manages language function; some left-handed patients may have semantic PPA from right temporal lobe degeneration.

Key Pathologic Features

Unlike nonfluent/agrammatic PPA, more patients with semantic PPA have had TDP-43, not abnormal tau as a dominant feature at autopsy. Some patients diagnosed with semantic PPA during life have had Alzheimer’s, not FTD changes in the brain at autopsy.

Genetics

The majority of semantic PPA cases are not hereditary.

Treatment

As with all forms of FTD, there is no cure for semantic PPA, and in most cases its progression cannot be slowed. Although no medications have been proven effective specifically in FTD, many clinicians look to the medications and treatment approaches targeting behavioral disturbances as necessary.

For instance, some FTD patients benefit from selective serotonin reuptake inhibitors (SSRIs, used in treating obsessive-compulsive behaviors, such as hoarding or craving sweets).

Management and Prognosis

Semantic PPA is a progressive disorder of language. In later stages of the disorder, some semantic PPA patients will develop symptoms common to the other FTD subtypes, including behavioral, social or motor difficulties. The language difficulty requires education for caregivers on the misunderstood nouns and gentle distraction of the agitated uncomprehending patient. Average duration of illness is 8-10 years. Parkinson-like immobility and swallowing difficulties may occur and lead to the end of life.

Reference: Gorno-Tempini, M.L., Hillis, A.E., Weintraub, S, et.al. Classification of primary progressive aphasia and its variants. *Neurology*; March, 2011.

For additional information and support:

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