aphasia
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Diagnosing Primary Progressive Aphasia (PPA)

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Aphasia is the loss or impairment of the use of language due to brain damage. The signs of aphasia are errors in verbal output including word-finding problems, trouble understanding speech, and difficulty in reading and/or writing.

Language is a "localized" brain function, meaning that some brain regions are more important for language than others. (In contrast, personality is mostly a non-localized brain function.) Aphasia usually results from damage to the left side of the brain; it is unusual for aphasia to result from right-sided brain damage. The severity of aphasia and prognosis for recovery depend on the volume of tissue damage and its exact location(s), the type of injury, and whether other parts of the brain are also damaged. The onset of aphasia is usually sudden as the cause is most often stroke or traumatic brain injury. Infection can also cause the rapid onset of aphasia.

The onset of aphasia is, however, not always sudden. "Primary Progressive Aphasia" (PPA) is one of several labels attached to a language disorder "of insidious onset, gradual progression and prolonged course, in the absence of generalized cognitive impairments (at least for a substantial period of time), due to a degenerative [brain] condtion" (Duffy). In layperson's terms, it is a slowly worsening aphasia not due to stroke, trauma, tumor or infection. It is sometimes called "progressive aphasia without dementia." Mesulam, who coined the term "primary progressive aphasia," believes that the aphasia must exist with no significant memory, behavioral, intellectual or visual impairments at least for two years before one can correctly use it diagnostically. (In some cases these other symptoms do appear later on, in which case the diagnosis is amended.)

The type or pattern of the language disorder may differ from patient to patient. As with aphasia secondary to stroke, the manifestations depend on what parts of the left hemisphere are relatively more damaged at any given point in the illness. 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 (it is an effort for the person to speak and he or she produces fewer words). 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.

The average age of onset is 60; most people with PPA are between the ages of about 40 to 80. Men are affected twice as often as women. About half of PPA patients have a family history of dementia in a parent or sibling, indicating the existence of a genetic component. Half of all people with PPA will eventually develop cognitive or behavioral problems consistent with a more pervasive dementia syndrome, such as Alzheimer's disease or Fronto-Temporal-Limbic Dementia, after an average of five years. In other people, aphasia may remain relatively isolated or even be the sole manifestation for as long as 15 years. In general, the longer the duration of aphasia as an isolated symptom, the less likely that other signs of dementia will develop.

As with other "degenerative" brain diseases, including Alzheimer's disease, there are no totally reliable non-invasive diagnostic tests for PPA. Whether-or-not the aphasia is "pure", that is, unaccompanied by other cognitive impairments, may in some cases be difficult to establish through psychological assessments because aphasia can affect

performance on “non-language “ (e.g., memory) portions of the test. CT scans, MRI, EEG (brain wave) often suggest left hemispheric damage, but these tests may also be normal even though the person has PPA. “Metabolic” imaging studies such as single photon emission computed tomography (SPECT), positron emission tomography (PET), and Functional MRI indicate left hemispheric dysfunction when a person has PPA, but other disorders with different symptoms may also cause the same radiologic abnormalities.

The cause of PPA and other degenerative brain disorders is unknown. A variety of brain abnormalities in people with PPA have been seen during autopsies. These abnormalities have involved the left hemisphere either exclusively or to a much greater extent than the right hemisphere. Most often, brain abnormalities are consistent with Fronto-Temporal-Limbic Dementia (FTLD), itself a syndrome--not a single disease--with variable symptoms and microscopic abnormalities. Although FTLD is usually associated initially with progressive changes in personality, sometimes it presents as PPA, personality deterioration occurring years later. Alzheimer’s disease has been proven the cause of PPA in a minority of patients. People with PPA due to Alzheimer’s disease usually have fluent aphasia.

Although researchers are testing different medications, as with other degenerative brain afflictions such as Alzheimer's, there is currently no cure for PPA. Speech/language therapy aimed at developing compensatory communication strategies may be useful for mildly impaired, motivated patients. These strategies may be the most helpful for those whose occupations are not strongly dependent on verbal communication.♦

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