Subtype: Nonfluent / Agrammatic Variant Primary Progressive Aphasia

The presenting feature in people with nonfluent/agrammatic PPA is deterioration in their ability to produce speech. These patients first become hesitant in their speech, begin to talk less, and eventually become mute. Current research suggests that the fundamental loss in nonfluent/agrammatic PPA is deterioration in knowledge of the grammatical organization and the production of sounds for language.

Unlike other FTD subtypes, nonfluent/agrammatic 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 aphasia in nonfluent/agrammatic PPA is experienced as hesitant, effortful speech. Despite this difficulty, it appears that patients' ability to comprehend what others say is preserved longer, though this is eventually lost, as well.

Increased difficulty producing speech due to weakness or incoordination – speech sounds weak, imprecise and uncoordinated.

Reading and writing abilities may be preserved longer than speech, but these eventually decline, as well.

Mutism eventually develops with progression.

Difficulty swallowing may develop late in the course of illness.

Neuroimaging studies demonstrate loss of brain volume in the left frontal and parietal areas on an MRI scan and/or decreased neural activity and blood flow, especially in the left frontal lobe on functional imaging (e.g., PET or SPECT scan).

In later stages, clinical features may include ones found more commonly in other FTD subtypes, particularly extrapyramidal syndromes such as corticobasal syndrome (CBS) and progressive supranuclear palsy (PSP).

Key Pathologic Features

Pathology in PPA is most commonly abnormal tau collections (FTLD-T). This abnormality in tau differs from that seen in Alzheimer's disease. We especially anticipate abnormal tau in cases of corticobasal syndrome, Pick's disease or progressive supranuclear palsy. Other cases of PPA may harbor TDP-43 deposits (a different type of protein abnormality from tau).

Genetics

Nonfluent/agrammatic PPA can be sporadic, familial, or hereditary. The majority of cases are not hereditary.

Treatment

As with all forms of FTD, there is no cure for PPA, and in most cases its progression cannot be slowed. Physicians suggest targeting behavioral disturbances as necessary (e.g., obsessive-compulsive behaviors, such as hoarding or craving sweets). Some physicians will give a trial of amantadine to ease the flow of speech, but this use has not been proven yet in a formal placebo-controlled drug trial. In one placebo-controlled randomized study, the cholinesterase inhibitor Reminyl (galantamine), significantly slowed deterioration and in some cases improved language function.

Management and Prognosis

Although no studies have shown improvement or slowing of progression, when a patient works with a speech and language pathologist (SLP), many centers work with SLPs to hone the diagnosis of PNFA or semantic dementia and to research potential therapeutic interventions.

Many PPA patients develop the behavioral, social and/or motor complications seen in other forms of FTD. In these patients, prognosis is obviously poorer and management more complicated. Patients who do not develop these additional symptoms are able to preserve their independence and active lifestyle for a longer period of time.

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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