



Nonfluent Variant Primary Progressive Aphasia

People with nonfluent variant primary progressive aphasia (nfvPPA) tend to come to the doctor's office with complaints about pronouncing words or increasing trouble getting words out. Their speech may sound slurred, or their voice may change. As time goes on, people with nfvPPA have more trouble putting sentences together, and they eventually begin to speak slower and slower.

Because it primarily affects the front part of the brain, nfvPPA is considered a subtype of a large group of brain conditions called [frontotemporal dementia](#) (FTD).

What Causes nfvPPA?

The cause of nfvPPA is unknown. Scientists know that in nfvPPA there is a build up of one of two proteins, either tau or TDP-43, usually in the front left part of the brain, which controls speech and language. These proteins occur normally, but we do not yet understand why they build up in large amounts in this brain area. As more and more protein builds up in those brain cells, the cells lose their ability to function and eventually die. This causes the brain to shrink.

How is Age Related to nfvPPA?

Most people with nfvPPA start to have symptoms in their 60s, although some people have shown signs earlier or later.

What Happens in nfvPPA?

People with nfvPPA gradually have more trouble expressing themselves, even though they still understand the meaning of words. They might begin speaking in shorter phrases or pausing mid-sentence or mid-word. They may have increasing difficulty with pronouncing or mixing up sounds in familiar words. People with nfvPPA may have trouble understanding sentences that are long or grammatically complex, and it may be hard for them to use correct grammar. Other

people may have a hard time understanding what the person with nfvPPA is trying to say. Speaking on the phone or in groups of people may be particularly difficult. Over time, people with nfvPPA may begin to use short sentences, omitting smaller words, like articles and adjectives. Reading and writing skills usually remain good in people with nfvPPA, so you might consider using a board or number of pictures to help the person express their meaning. Skills with numbers, colors and shapes generally remain intact, as do skills involving face and emotion recognition.

In advanced nfvPPA, people may have such difficulty saying words out loud that they might stop speaking completely, and they may become depressed, anxious or indifferent. Difficulties with planning and judgment may also happen. People with nfvPPA may have trouble with movement, especially on the right side of the body, such as difficulty moving the right arm or leg. People with nfvPPA may also have difficulty swallowing, which could become serious enough to interfere with eating and puts them at risk for infections like pneumonia.

nfvPPA is a disease that changes with time. A person with nfvPPA can live many years with the disease. Research has shown that a person with nfvPPA may live an average of 8–10 years with the disease, although this can vary from person to person.

Many patients with the nonfluent variant go on to develop parkinsonian symptoms that overlap with [progressive supranuclear palsy](#) (PSP) and [corticobasal syndrome](#) (CBS), such as an inability to move the eyes side-to-side, muscle rigidity in the arms and legs, and weakness in the muscles around the throat. People with nfvPPA tend not to show the behavioral characteristics of [FTD](#) until quite late in the disease, and they are keenly aware of their difficulties. Depression and social withdrawal are common features of nfvPPA. As the disease progresses, less and less language is used, as speech production becomes increasingly difficult. Changes to the brains of people with nfvPPA are seen in the left posterior frontal lobe and surrounding brain areas. The nonfluent PPA variant accounts for about 20% of all people with frontotemporal dementia.

Are There Medicines to Treat nfvPPA?

Though there is no cure for nfvPPA yet, there are medications that may help manage the symptoms. These medications are called selective serotonin reuptake inhibitors (SSRIs), and these medicines can help if a person with nfvPPA is having changes in mood or behavior.

What Other Things Help?

There are various ways to help a person with nfvPPA. Speech therapy may help improve communication between people with nfvPPA and others. Communication devices, like tablets and type-to-talk instruments, or picture books, can help the person with nfvPPA express what

they want to say with less anxiety.

If symptoms of weakness or difficulty using the arm or leg develop, physical and occupational therapy may help manage them and provide coping skills and strategies. A swallowing evaluation and a modification of diet and consistency of liquids may be necessary for people with nfvPPA who develop swallowing difficulties.

Research has shown that physical exercise helps to enhance brain health and improves mood and general fitness. A balanced diet, enough sleep, and limited alcohol intake are other important ways to promote good brain health. Other illnesses that affect the brain, such as diabetes, high blood pressure, and high cholesterol, should also be treated if present.

Resources

- [2011 Gorno-Tempini diagnostic criteria for PPA](#)
- [International PPA Connection](#)
- [National Aphasia Association](#)
- [Faces of Aphasia](#)
- [Aphasia Hope Foundation](#)
- [The Association for Frontotemporal Degeneration](#)
- [American Stroke Association](#)
- [Family Caregiver Alliance](#)
- [National Institutes of Health](#)

Participate in Research

- [Clinical trials at UCSF](#)
- [ClinicalTrials.gov](#)