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Patient Education

Primary progressive aphasia

An overview of primary progressive aphasia

Primary progressive aphasia is a neurodegenerative disorder. Neurodegenerative means that certain nerve cells in the brain have been damaged or no longer work. This condition is called PPA.

Primary progressive aphasia is a gradual loss of language skills. It starts slowly and progresses over time. Other speaking and thinking skills are mostly intact early on. Over time, they may be affected too.

Aphasia

Aphasia is a condition that makes it hard to use words, understand words or both. This happens when someone talks, listens, reads and writes.

Aphasia may affect four different parts of language:

- **Sounds.** People with aphasia may have trouble with the sounds that make up words. For example, someone with aphasia may say bat when they meant to say pat. The study of sounds is called phonology.
- **Word markers.** These are the small parts of words that change their meaning. For example, when you add an "s" to dog, it means there is more than one dog. Another example is to add "ed" to a verb. That ending means something happened in the past. Someone with aphasia might leave these word markers out or use them incorrectly. For example, they may say walk instead of walked. The study of word markers is called morphology.
- **Word meaning.** Aphasia can make it hard to find the right words. For example, people may say fork when they mean to say spoon. Aphasia also can make it hard to understand what words mean when other people say them. Some people with aphasia may not remember what a word means. You may hear word meaning called semantics.
- **Grammar.** It can be hard for a person with aphasia to put words in the right order to make a sentence. For example, instead of saying, "The dog is chasing the cat," someone with aphasia may say, "Dog cat chasing." You may hear grammar called word order or syntax.

Aphasia affects these key building blocks of language. This can make communication hard for people and those around them.

It is important to know that **aphasia does not affect intelligence. People with aphasia have typical thoughts, ideas and feelings.** But they may need your patience and support to express them.

Types and symptoms of primary progressive aphasia

There are three main types of primary progressive aphasia. Types also are called variants. Each one is based on the part of the language area that is affected.

- **Sounds.** Logopenic variant primary progressive aphasia.
- **Words.** Semantic variant primary progressive aphasia.
- **Grammar.** Nonfluent or agrammatic variant primary progressive aphasia.

Each type of primary progressive aphasia causes different symptoms at the start. Other symptoms develop over time. They gradually worsen.

Many people say that the later symptoms of PPA are less severe than the difficulty they had with early symptoms. But those later symptoms may have an equal impact on quality of life. Or they may have an even greater impact on a person's life. For example, changes in swallowing may be less severe, but they may be very challenging.

Symptoms of logopenic variant primary progressive aphasia

People with the logopenic variant:

- Have difficulty thinking of the words they want to say.
- Often try to describe the word they are missing.
- May use the wrong word, especially one that sounds alike, but isn't quite right.
- Have trouble with reading and spelling. For example, reading or writing pother instead of bother.

As this condition progresses, other symptoms may emerge, such as:

- Finding it difficult to remember, think or solve problems.

This variant typically is caused by the proteins amyloid and tau in the brain. Those proteins also are associated with Alzheimer's disease.

Symptoms of semantic variant primary progressive aphasia

People with the semantic variant lose their understanding of the meaning of words.

When they talk:

- They may use words that are not specific, such as thing.
- They may use words that are not correct but are related to the word they want to say. For example, they may call all animals a dog.
- They may use a very general word instead of a specific word. For example, a person may use woman instead of daughter.
- They may have trouble with spelling. Often, this first happens with words that are not spelled as they sound. One example is writing yot for yacht.

When they listen:

- They may not recognize or understand words they used to know.

As this variant progresses, other symptoms may appear, such as:

- Trouble with breaks in a routine.
- Fixation on certain activities, such as puzzles.
- Having a sweet tooth.
- Finding it difficult to recognize people's faces.

This variant typically is caused by a protein in the brain called TDP. This is a short form for TAR DNA-binding protein.

Symptoms of nonfluent or agrammatic variant primary progressive aphasia

Nonfluent refers to speech that takes a lot of effort to form. Sometimes, it refers to using words out of order. Agrammatic refers to using words in a way that is not typical English grammar. It's common for people with this variant also to have apraxia of speech. That condition makes it hard for a person to speak clearly or quickly.

With nonfluent or agrammatic primary progressive aphasia, people:

- May not speak in complete sentences. They may have trouble putting words in the right order. They may leave out small connecting words, such as the word the. For example, they may say, "Girl eat cookie."
- May say yes when they mean to say no.
- May have difficulty understanding information that is shared in a complex way.

This variant can be caused by different proteins, depending on the mix of speech and language problems a person has.

- When apraxia of speech is present, nonfluent or agrammatic primary progressive aphasia symptoms typically are caused by tau proteins. This is particularly true for corticobasal degeneration or progressive supranuclear palsy. See also "Apraxia of speech."
- When nonfluent or agrammatic primary progressive aphasia is present but the person does not have apraxia of speech, the condition may be caused by a type of tau protein that is associated with Pick bodies.

As this variant progresses, other symptoms may appear. Examples include:

- Trouble with sound sequencing or planning. This is called apraxia of speech.
- Trouble with the physical act of speaking. This may be muscle weakness or muscle tightness. This is called dysarthria.
- Trouble swallowing. This is called dysphagia.
- Tightening of a group of muscles. This is called spasticity or rigidity.
- Trouble with movements such as slowness or walking problems.

Primary progressive aphasia can be mixed

Some symptoms of primary progressive aphasia may not fit clearly into one of the three main types. This often is true with a later diagnosis. This may be called a mixed type or a type that cannot be classified.

The need for regular checkups

Each person with primary progressive aphasia is different. It's hard to predict whether new symptoms may develop. And it's hard to predict when that may happen. Talk with your care team about your schedule for regular checkups. Be sure to tell the care team about any new or worsening symptoms you notice.

Apraxia of speech

Apraxia of speech is a separate condition. It may occur along with aphasia. It often happens with the nonfluent or agrammatic type of aphasia. It can occur without aphasia.

Apraxia of speech affects the ability to speak clearly or quickly. But it does not affect the ability to understand others or to read or write.

With apraxia of speech, the brain struggles to make the lips, jaw and tongue move correctly. A person finds the words to say, but the speech muscles do not move correctly to make the needed sounds or words. The speech that comes out may be slow or sound jumbled.

With apraxia of speech, there may be issues with:

- **Sequencing.** This is the order of sounds or syllables as they are spoken.
- **Prosody.** This is the speed and animation with which a person speaks.
- **Articulation.** This refers to how words are formed.

When apraxia of speech occurs by itself

Apraxia of speech may occur by itself. It may have no connection to primary progressive aphasia. When it occurs alone, often it is called primary progressive apraxia of speech. You may see or hear it called PPAOS.

Primary progressive aphasia and dementia

Dementia refers to conditions in which a person's memory, thinking and social abilities affect daily activities. Two common causes of dementia are Alzheimer's disease and frontotemporal dementia.

A person can be diagnosed with primary progressive aphasia and still be able to live alone with no support. **But when people have difficulty caring for themselves, they may be diagnosed with dementia.** Examples of self-care activities are dressing, bathing, driving and taking care of money.

As primary progressive aphasia progresses, signs of dementia may be identified. At that time, a diagnosis of both aphasia and dementia may be made.

Causes of primary progressive aphasia

Primary progressive aphasia happens when certain parts of the brain shrink. These parts are called lobes. In this case, the frontal, temporal or parietal lobes are affected. It is possible that all of them may be affected.

When areas of the brain shrink, it's called atrophy. The atrophy caused by primary progressive aphasia mainly occurs on the left side of the brain. These areas are responsible for speech and language. The right side of the brain often is involved, too, but to a lesser degree. It may not be involved until later in the disease.

Atrophy is associated with the presence of certain proteins in the brain. The proteins are believed to lessen brain activity or function. The proteins amyloid, tau and TDP often are related to primary progressive aphasia. See "Types and symptoms of primary progressive aphasia" for more information about each type.

Most people diagnosed with PPA do not have a family history of similar conditions. This is called a sporadic occurrence. However, genetic factors may play a role. This is especially true for some variants. Be sure to tell your care team if any family members have had similar changes.

Diagnosing primary progressive aphasia

It may be hard to diagnose this condition. Symptoms can be confused with other conditions. It is important to be honest with your care team about any symptoms you notice. The information you share can make it easier to diagnose any condition you have.

When you have symptoms of PPA, your primary care team may refer you to other specialists. For example, you may see:

- **A neurologist.** This specialist diagnoses and treats disorders of the brain, spinal cord and nervous system.
- **A behavioral neurologist.** This specialist works with people who have brain conditions that affect memory and thinking.
- **A speech-language pathologist.** This specialist helps diagnose and treat a wide range of speech, language, thinking and swallowing conditions. Speech-language pathologists work with you to identify your communication challenges and strengths. Together, you create a treatment plan to fit your needs.

Additional testing

Your healthcare team may refer you for neuropsychological testing. It examines memory, reasoning and judgment. It examines problem-solving ability, language skills and visuospatial abilities. Visuospatial abilities refer to the way you relate visual information to the space around you.

Your care team may advise you to have a PET scan. It is an imaging test that uses radioactive medicine called a radioactive tracer. The test creates multiple images of the brain. PET stands for positron emission tomography.

There are several different types of PET scans. An FDG PET scan is the most sensitive brain test used to diagnose primary progressive aphasia. This scan looks at where the brain processes sugar. **Specifically, it shows the areas of the brain that do not process sugar.** The disease often is found in those areas.

FDG is the short name for fluorodeoxyglucose. It is a radioactive tracer used in the FDG PET scan. The amount of medical-grade radioactivity used is as low as possible to find and show metabolic activity in the body.

Treatment of primary progressive aphasia

There is no cure for PPA at this time. But treatment can ease the effect of symptoms. It also can be used to slow the progression of symptoms.

Each person with this condition is different. Treatment depends on your symptoms and their severity. It depends on how the condition progresses. And it depends on your priorities and goals.

Possible treatment options include:

- **Speech-language therapy.** This helps maintain your communication abilities. It also allows you to adapt to new changes and to prepare for future changes. Therapy can help you explore other ways to communicate if necessary. And it can help with swallowing difficulties if they develop.
- **Physical and occupational therapy.** Therapy specialists help you manage movement problems.
- **Counseling.** These specialists help you manage anxiety and depression.
- **Palliative care.** These specialists help you think through the diagnosis. They also work with you to plan how to live your life with a progressive illness.

If Alzheimer's disease is thought to be the cause of your PPA, medicine may be a treatment option. Talk to your care team about this if needed.

Your healthcare team works with you to decide the best therapies for your symptoms.

Common questions about primary progressive aphasia

Is there any research going on for primary progressive aphasia?

Research for PPA is ongoing. If you meet research study guidelines, you may be invited to take part. This decision is up to you. Your decision does not affect your care.

If you decide to take part in a research study, be sure you understand:

- The purpose of the study.
- Any risks the study may have.
- How long the study lasts.
- Whether you can leave the study at any time.
- What you are expected to do as a part of the study.

What are some tips to communicate with a person who has primary progressive aphasia?

- **Be patient and give extra time.** Allow people with PPA time to express themselves. Do not rush or interrupt them.
- **Simplify your language.** Use short sentences and simple words. When possible, ask questions that need only yes or no answers or brief responses.
- **Use gestures and visual aids.** If you need to clarify your message, point, draw or show pictures while you talk. Visual cues help support understanding.
- **Limit distractions.** Lessen background noise and other distractions. Give all your attention to someone.
- **Encourage nonverbal communication.** If speaking is difficult, support the use of gestures, writing or communication apps.
- **Do not pretend to understand.** If you don't understand, be kind and honest.

What are some tips for a person who has primary progressive aphasia?

- Take time and describe words if you can't find them.
- Use gestures, pointing or writing in addition to speaking.
- Keep communication tools such as apps or writing boards close by.
- Plan key points before important conversations.
- Work with a speech-language pathologist to develop strategies.
- Advocate for your needs. Tell other people how to help you in different communication settings. For example, you may want certain help at home. But you may want different types of help when you are in public with friends.

Need more information?

If you have questions after you read this, talk with a member of your healthcare team. Each member of your care team is here to help you.

For more information, see these related websites:

- National Aphasia Association.
- The Association for Frontotemporal Degeneration.
- CurePSP (progressive supranuclear palsy).
- Alzheimer’s Association.
- FTD Disorders Registry (frontotemporal degeneration).
- American Speech-Language-Hearing Association (ASHA).
- U.S. National Library of Medicine, clinical trials.

Notes

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Barbara Woodward Lips
PATIENT EDUCATION CENTER

Mrs. Lips, a resident of San Antonio, Texas, was a loyal patient of Mayo Clinic for more than 40 years. She was a self-made business leader who significantly expanded her family's activities in oil, gas and ranching, even as she assembled a museum-quality collection of antiques and fine art. She was best known by Mayo staff for her patient advocacy and support. Upon her death in 1995, Mrs. Lips paid the ultimate compliment by leaving her entire estate to Mayo Clinic.

Mrs. Lips had a profound appreciation for the care she received at Mayo Clinic. By naming the Barbara Woodward Lips Patient Education Center, Mayo Clinic honors her generosity, her love of learning, her belief in patient empowerment, and her dedication to high-quality care.

> This information is for your education only. It does not replace medical advice, diagnosis or treatment. New medical research or practices may change this information. If you have questions about a medical condition, talk with a member of your healthcare team.

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