



A Healthcare Provider's Guide To The Logopenic Variant Of Primary Progressive Aphasia (lvPPA):

Diagnosis, pharmacologic management,
non-pharmacologic management, and
other considerations

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Diagnosis

Definition

Primary progressive aphasia (PPA) is diagnosed when patients present with progressive decline in language functions as a predominant feature. There are three types of PPA, the logopenic variant (lvPPA) is one of them. Patients with lvPPA have difficulties with the phonological processing of language.

Etiology

The cause of lvPPA is unknown. In most patients, pathology from autopsy reveals the presence of plaques and tangles similar to that seen in classical Alzheimer's disease. Brain Magnetic resonance imaging (MRI) in lvPPA usually reveals parenchymal volume loss in the left angular gyrus and left temporoparietal junction, both involved in auditory and phonological processing.

Course

Patients with lvPPA have difficulty with phonological processing of language. They often present with word finding difficulty and at times complain that they can't "hear" other people. Because of that, they are often not able to hold on to short-term information, such as when given an instruction to follow or a phone number to dial. Because of that, they may often complain that they have trouble with memory. They may have more difficulties in loud and crowded places where phonological processing is key to distinguish speech from noise. Patients with lvPPA may have a slow and hesitant speech with many word finding pauses. They have difficulties repeating sentences. Patients with lvPPA may also have difficulty with calculation or memory tasks.

As the disease progresses, patients may lose their speech completely and become mute. They may also develop more classical Alzheimer's disease symptoms such as memory loss, difficulty finding their way or recognizing familiar faces. The

disease may progress to affect motor skills and swallowing in more advanced stages.

The logopenic variant of primary progressive aphasia needs to be differentiated from the other types of PPA, especially the non-fluent variant (nfvPPA) as they have many overlapping symptoms but distinct pathological substrates. Other types of dementia such as the classical amnesic syndrome of Alzheimer's disease should also be ruled out. A strategically located stroke or brain tumor may mimic symptoms of lvPPA, especially if the presentation is more acute. Finally, as with other types of dementia, a work-up should rule out potentially reversible metabolic, infectious and inflammatory causes of cognitive decline.

Diagnostic Criteria

The following are the diagnostic criteria for primary progressive aphasia in general and for the logopenic variant more specifically:¹

Primary Progressive Aphasia:

- I. Inclusion Criteria (All of a-c must be present)
 - A. Most prominent clinical feature is difficulty with language
 - B. Deficits are the principal cause of impaired daily living activities
 - C. Aphasia should be the most prominent deficit at symptom onset and for the initial phases of the disease
- II. Exclusion Criteria
 - A. Pattern of deficits is better accounted for by other non-degenerative nervous system or medical disorders
 - B. Cognitive disturbance is better accounted for by a psychiatric diagnosis
 - C. Prominent initial episodic memory, visual memory and visuoperceptual impairments
 - D. Prominent initial behavioral disturbances

Diagnostic Criteria for Logopenic Variant PPA

- I. Clinical diagnosis of LV-PPA
 - A. Impaired single-word retrieval in spontaneous speech
 - B. Impaired repetition of sentences and phrases
 - C. Speech (phonological) errors in spontaneous speech and naming
 - D. Spared single word comprehension and object knowledge
 - E. Spared motor speech (no distortions)
 - F. Absence of frank agrammatism
- II. Imaging supported diagnosis of LV-PPA
 - A. Clinical diagnosis of LV-PPA
 - B. Imaging must show one of:
 - a. Predominant left posterior perisylvian or parietal atrophy on MRI
 - b. Predominant left posterior perisylvian or parietal hypoperfusion or hypometabolism on Single-photon emission computed tomography (SPECT) or Positron Emission Tomography (PET)

Pharmacologic Management

Medications to Use

There is no known cure for lvPPA. Because the underlying pathology is most commonly Alzheimer's disease, many providers use cholinesterase inhibitors such as donepezil or rivastigmine similarly to how these medications are used in classical Alzheimer's disease.

If symptoms of depression or anxiety are present in people with lvPPA, selective serotonin reuptake inhibitors (SSRI) may be used as treatment.

Medications to Avoid

Medications with strong anticholinergic side effects, such as sedating antihistamines, barbiturates, narcotics, benzodiazepines, gastrointestinal and urinary antispasmodics, central nervous system (CNS) stimulants, muscle relaxants, and tricyclic antidepressants should be avoided. Antipsychotics should be used with caution. If used, carefully evaluate effectiveness of medication and consider discontinuing if there is no improvement in six weeks.^{2,3,4}

Non Pharmacologic Management

Speech Therapy

There is some preliminary evidence supporting the benefit of speech therapy. For example, speech and language therapy may slow the decline in naming abilities.^{5,6} In a small group of patients with left-hemisphere lesions, singing promoted word intelligibility.⁷

Healthy Lifestyle

There are lifestyle habits that promote health and well-being. Research suggests that the combination of good nutrition, physical activity, and mental and social engagement may provide benefit in promoting health although more study is needed to determine the actual mechanisms.^{8,9} A heart-healthy diet (lower in sugar and fat and higher in vegetables and fruit) is considered good for both the body and the brain. An example is the Mediterranean diet that promotes nutrition based on fruit, vegetables, nuts, and grains with limits on consumption of red meat and saturated fats. Physical exercise has been associated with improvement of mood and mobility, and a decrease in the risk for falls.^{10,11} Physical activities that are socially engaging (walking or swimming with a friend and participating in exercise groups) can be especially enjoyable. Engagement in activities that are mentally stimulating (crossword puzzles, sudoku, computer games) is encouraged as long as the activity is enjoyable.

The Alzheimer's Association has more information on tips for maintaining your health: www.alz.org/we-can-help-brain-health-maintain-your-brain.asp

Sleep

Disrupted sleep can negatively impact memory and thinking, though the mechanisms are not well understood.¹²

Components of sleep hygiene include:

- Avoid napping during the day
- Avoid stimulants such as caffeine, nicotine, and alcohol too close to bedtime
- Get regular exercise
- Avoid eating right before sleep
- Ensure adequate exposure to natural light
- Establish a regular relaxing bedtime routine
- Associate your bed with sleep. It's not a good idea to use your bed to watch TV, listen to the radio, or read.

For more details on sleep hygiene, you can refer to the National Sleep Foundation at <http://sleepfoundation.org/ask-the-expert/sleep-hygiene>.

Other Considerations

Support Resources

- Alzheimer's Association: alz.org
- Family Caregiver Alliance: caregiver.org
- National Institute of Health/National Institute on Aging: nia.nih.gov/alzheimers
- The Association for Frontotemporal Degeneration: theaftd.org

- National Aphasia Association: aphasia.org
- The Primary Progressive Aphasia Program at Northwestern University: brain.northwestern.edu/dementia/ppa/index.html
- Primary Progressive Aphasia support, an online support forum: groups.yahoo.com/neo/groups/PPA-support/info

Research and Clinical Trials

The National Institutes of Health maintains an extensive listing of clinical trials at www.clinicaltrials.gov. Academic medical centers may be engaged in research and clinical trials.

Safety

Patients with language disorders may have difficulties being left home alone in case there is an emergency and they need to call 911, however are unable to explain the situation because of language problems. Caution is advised and sometimes the early introduction of a companion is helpful.

Driving

Though in the early stages of the disease, patients with lvPPA may not have difficulty operating a car, they may progress to having more difficulty finding their way. Also, because of their speech difficulties, if they are stopped by the police, they may be thought of as inebriated. A letter from the physician explaining the clinical diagnosis and kept in the car may be helpful. A driving evaluation with the Department of Motor Vehicles (DMV) is often recommended.

Depending on cognitive and motor findings, the patient may be requested to stop driving, complete test of driving abilities through the DMV, or be referred to a driver's safety course that will assess driving ability. Reporting to the department of motor vehicles should be consistent with state laws. Some states have mandatory reporting requirements: the diagnosis is reported to local health departments who then report to the DMV. Individual state requirements can be found at: dmvusa.com.

Living Situation and Environment

It is important to determine if the patient's residential setting best meets his or her functional and cognitive abilities. Areas of concern may include personal safety (ability to manage medications safely,

ability to manage nutritional requirements, ability to manage personal hygiene) and quality of life (activities and engagement that match the person's needs and abilities).

Types of living situations range from living at home alone or living at home with supervision, to board and care, assisted living, or memory care units.

Elder Abuse

Patients with dementia and their caregivers are vulnerable to abuse. Refer to Adult Protective Services (APS) if there is concern for the well-being of the patient or the caregiver.

To locate an APS office in your state, see: napsa-now.org/get-help/help-in-your-area/.

Legal Planning

Provide information about advance directives and durable power of attorney while the patient is in the early stages of disease and able to articulate his or her wishes. Make referrals for legal and financial advice, especially if there are concerns about the patient's judgment, decision-making, or vulnerability. A formal evaluation for capacity may be warranted. The Alzheimer's Association provides a brochure that covers legal planning: alz.org/national/documents/brochure_legalplans.pdf.

- **Advanced Directives**
These documents allow individuals to state their preferences for medical treatments and to select an agent or person to make health care decisions in the event they are unable to do so or if they want someone else to make decisions for them.
- **Power Of Attorney**
A Power of Attorney (POA) is a legal document that gives someone of an individual's choosing the power to act in their place. POAs can be for medical or financial matters.
- **Living Will**
A living will is a written, legal document that spells out medical treatments that an individual would and would not want to be used to keep them alive, as well as other decisions such as pain management or organ donation.

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