

The SLP's Guide to PPA and PPAOS

What is Primary Progressive Aphasia (PPA)?

- A neurodegenerative condition that begins with progressive deterioration of language skills.
- Motor speech skills may or may not be affected.
- Other neural domains are relatively spared in the beginning.
- Eventually experience cognitive, motor, and behavioral impairments.

(Wauters et al., 2023)

What are the 3 variants of PPA?

- Nonfluent/agrammatic variant, semantic variant, and logopenic variant.
- The information below is from Wauters et al. (2023). See Duffy et al. (2015) for more info.

Nonfluent/agrammatic variant (nfvPPA)

- Grammatical errors or effortful/halting speech consistent with apraxia of speech.
- Dysarthria may or may not be present.
- Must have at least 2 of the following:
 - Impaired comprehension of syntax.
 - Intact single word comprehension.
 - Spared object knowledge.
- Neuroimaging: includes left posterior-frontal/insular atrophy.
- Most likely associated with FTLD-tau (frontotemporal lobar degeneration).
- May progress to include impairments of executive function and motor skills.

Semantic variant (svPPA)

- Impaired objects/picture naming.
- Deficits in single word comprehension.
- At least 3 of the following:
 - Impaired object knowledge.
 - Surface dyslexia or dysgraphia (deficits in reading or spelling irregular words).
 - Able to repeat.
 - Spared grammar.
 - Intact motor speech skills.
- Neuroimaging: Anterior temporal atrophy (left hemisphere > right).
- Most likely associated with TDP-43 inclusions.
- May progress to include deficits in sleep, appetite, and recognizing faces. May also experience altered libido, emotional blunting, dis-inhibition, and other behavioral changes.

Logopenic variant (lvPPA)

- Impaired word retrieval.
- Deficits in phrase/sentence repetition.
- At least 3 of the following:
 - Phonological errors in speech.
 - Grammar is intact.
 - Spared single word comprehension.
 - Intact motor speech skills.
- Neuroimaging: atrophy in left temporoparietal areas.
- Most likely associated with Alzheimer pathology.
- May progress to include episodic memory deficits, limb apraxia, and visuospatial deficits.

What is Primary Progressive Apraxia of Speech (PPAOS)?

- A progressive neurodegenerative condition that affects motor speech praxis, leaving language skills relatively intact. (Wauters et al., 2023)
- PPAOS can occur without aphasia or dysarthria. (Duffy et al., 2015).
- A relatively new diagnostic category. The definition of nfv(PPA) still includes people who have PPAOS without aphasia. See Duffy et al. (2015) for discussion.

What we know about PPAOS (Duffy et al., 2015; Wauters et al., 2023)

- Symptoms are similar to acquired AOS following a stroke.
 - Impaired rate: slow rate, increased pauses between syllables, lengthened sounds.
 - Deficits in repetition: distortions, additions, repetitions, and substitution.
- Neuroimaging: Includes involvement of the superior lateral premotor and supplementary motor cortices, as well as certain subcortical areas. (Duffy et al., 2015)
- Most likely associated with progressive supranuclear palsy or corticobasal degeneration (Duffy et al., 2015), with FTLT-tau indicated in well-characterized cases (Wauters et al., 2023).

Overview of Wauters et al. (2023) systematic review (open access)

- Speech therapy interventions for PPA and PPAOS.
- 45 higher-quality studies (304 participants) were identified.
 - 110 participants had nfvPPA.
 - 93 had svPPA.
 - 96 had lvPPA.
 - 4 had PPAos.
 - 1 had mixed PPA.
- 16 of 304 participants had AOS but none were identified as having PPAOS.
- 146 males, 155 females, 6 not reported.
- Mean age was 67.1 years.

Overview of treatments reported in the 45 higher-quality studies

- 37 studies: spoken naming or lexical retrieval.
- 4 studies: semantic knowledge.
- 3 studies for each: syntax/morphology, speech production/fluency, and written naming/spelling.
- 2 studies for each: discourse, functional communication, AAC/multimodal communication.
- 1 study: word comprehension.

Lexical retrieval included:

- Errorless learning.
- Cueing or self-cueing for word retrieval (such as phonological, orthographic, semantic, or autobiographical).
- Generative naming.
- Synonyms and antonyms.
- Conversational practice with a communication partner.

Other treatment approaches included:

- Script training.
- Constraint-induced aphasia therapy.
- Using AAC or other assistive device (ex. smartphone to search for target words for recipe).
- Written naming treatment.
- Training multimodal communication (ex. writing, gesturing).
- Explicit instruction of morphosyntactic structures.

Treatment outcomes for PPA

- Every study reported a positive outcome for at least one participant.
- 34 of 43 studies showed positive generalization for at least one participant.
- 34 of 38 studies showed maintenance of treatment gains for at least one participant.
- 17 of 19 studies reported a positive outcome on a social validity measure for at least one participant.

Post-hoc analysis of individual participants. Of the 45 higher-quality studies, 29 studies reported results for 56 participants.

- 53 of 56 participants improved on the primary outcome measure.
- 33 of 53 participants showed generalization.
- 39 of 46 participants maintained their treatment gains (for varying times).
- 17 of 18 participants reported improvement on a social validity measure.

Treatment outcomes for PPAOS

- None of the 45 higher-quality studies included treatment for people diagnosed with PPAOS.
- Five of the higher-quality studies did include 16 participants with AOS.
- Four studies included speech production as a target and reported positive results.

Assessing the intervention approaches for PPA

- As there are no randomized controlled trials, no intervention approach can be designated as a **practice standard**.
- Criteria met to be considered **practice guidelines**:
 - Lexical retrieval treatment: All subtypes of PPA.
 - Script training: nfvPPA.
- Criteria met to be considered **practice options**:
 - All other interventions included in the higher-quality studies: All subtypes of PPA.

Assessing the intervention approaches for PPAOS

- Evidence is lacking, although there is some evidence that behavioral therapy can be effective.
- Until we have more evidence, rely on the evidence for nondegenerative AOS, clinical experience, expert opinion, and general principles for treating other degenerative motor speech disorders.

Implications for clinical practice (WHO-ICF)

I'm adapting recommendations from Baylor & Darling-White's (2020) framework and Yorkston et al. (2017). We can assess and treat each area of the WHO-ICF model while focusing on a single communicative participation situation that is important to our patient.

Communicative Participation (Baylor & Darling-White, 2020)

- Find one single, specific situation our patient would like to succeed with communication. All of our therapy activities would revolve around this situation. If they have trouble selecting a specific conversation/situation, we can use:
 - Motivational Interviewing.
 - Life Interests and Values Cards (Haley et al., 2013).
 - Other tools, such as a needs assessment.
- Assess baseline and outcomes with a patient reported outcome measure (PROM)
 - If you can't find a relevant PROM that would capture your intended outcome, create your own - it's easy!
 - Self-anchored rating scale such as Likert scale, visual analog scale.
 - Goal attainment scaling (can be objective or subjective).
- Design intervention around this single situation.

- Use relevant words/phrases/sentences in your impairment-based treatments (word finding, scripts, etc).
- Teach and support implementation of strategies to counteract environmental or social barriers.
- Address personal perspectives related to the targeted conversational situation.

Impairment (body structure/function)

- Lexical retrieval treatments are impairment-based treatments (see above for the treatments used by the 45 higher-quality studies in the systematic review).
- Assess and treat as we usually do, except choose stimuli relevant to the communication participation situation our patient wants to improve.
- May be most effective early in the course of the disease.
 - Prophylactic treatment to slow the decline. (Wauters et al., 2023)
 - Impairment-based restorative treatment to regain lost skills. (Wauters et al., 2023)

Activity

- Facilitate our patient's ability to use lexical retrieval strategies in functional tasks.
- Script training.
- Using AAC or other supportive technology.

Physical and social factors (environment) (Baylor & Darling-White, 2020)

- Physical environment
 - Background noise, projecting voice over distance, speech-based technology, educational materials that are difficult to read or understand.
- Social environment
 - Attitudes, support, relationships.
 - Formal services and policies.
- Select a barrier that can reasonably be reduced.
- We can teach recommendations or brainstorm with our patient on ideas for what would work for them. We can assist our patient with implementing the strategy and brainstorm next steps.

Personal factors (Yorkston et al., 2017)

- Includes personal identity, life views, coping mechanisms, priorities, and values.
- We can provide education, training, coaching, and guidance on many aspects of self-management and self-efficacy skills related to communication and life participation.
- SLPs are experts at helping people improve the self-efficacy skill of performance mastery, but we can do so much more!
 - Self-management skills: problem-solving, decision-making, accessing and using resources, relationships, and taking action.

- Self-efficacy skills: performance mastery, finding peer models, reinterpreting symptoms, and social persuasion.
- Assess with motivational interviewing, PROM, needs assessment, self-anchored rating scales, or collaborative GAS.
- Key to remember: negative emotions and self-limitations do not correlate with degree of impairment.
- Skilled intervention ideas include:
 - Teaching our patients how to problem-solve their own participation-level problems.
 - Training and coaching our patients in the use of strategies and self-advocacy.
 - Teaching our patients about their condition and what to expect.
 - Sharing resources about support groups, conversation groups, literature, media by other people who have the condition, or participating in research.
 - Assisting patients to access and use resources.
 - Teach strategies and assist our patients in preparing for medical treatments.
 - Devising action plans that feel doable to our patient.
 - Exploring the possibility of re-interpreting symptoms. For example, it is likely that external factors contribute to our patient's communication difficulties, but many patients may assume that the burden is fully on themselves to change or deal with it.

Learn more

- Sarah Baar interviews Lisa Wauters, first author on the systematic review on PPA/PPAOS: <https://www.medbridge.com/course-catalog/details/is-there-reason-to-hope-with-PPA-sarah-baar-lisa-wauters/>
- Open access systematic review: <https://link.springer.com/article/10.1007/s11065-023-09607-1>
- How to write participation level goals with Baylor & Darling-White's (2020) model: <https://eatspeakthink.com/participation-level-speech-therapy-goals/>

Selected References

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