Nonfluent Agrammatic Primary Progressive Aphasia (nfaPPA)

Dementia Neuropsychology · https://neuro.vip/neuropsychology/dementia-neuropsychology/nonfluent-agrammatic-primary-progressive-aphasia-nfappa/

Nonfluent/agrammatic primary progressive aphasia (nfaPPA), sometimes referred to as the nonfluent variant PPA, is a neurodegenerative disorder prominently affecting the **expressive language system**. Unlike language loss caused by stroke or trauma, nfaPPA is characterised by a **slow, relentless impairment of speech production, grammar, and (frequently) speech motor planning**, while most other cognitive domains remain comparatively preserved in the early course. For clinical neuropsychologists, distinguishing nfaPPA from other language and cognitive disorders is vital for diagnosis, management, neuropsychological rehabilitation and support planning.

Clinical Presentation in Nonfluent Agrammatic Primary Progressive Aphasia

Core Language Features

Individuals with nfaPPA usually present with a combination of the following symptoms:

Effortful, Halting Speech

Speech is typically slow, laboured, and dysfluent. Patients may struggle to “get words out,” producing short phrases or single words rather than full sentences. The overall speech output is noticeably reduced compared with their premorbid abilities.

Agrammatism

There is a marked deterioration in grammatical construction (syntax) during both speech and writing. Sentences become “telegraphic,” often omitting function words (such as ‘is’, ‘the’, or ‘of’) and verb inflections. For example, “She go shop yesterday” instead of “She went to the shop yesterday.”

Speech Apraxia

Motor speech difficulties, including apraxia of speech, are strongly associated with nfaPPA. Patients may produce distorted sounds, dysprosody (altered rhythm and stress), or sequencing errors (e.g. “tup” for “cup”) unrelated to muscle weakness (dysarthria).

Confrontation Naming

Word-finding difficulties may be present, but are generally **less prominent than in other primary progressive aphasia variants**. When naming objects, patients may substitute phonologically similar words or produce sound-level errors due to apraxia:

Phonologically similar word substitutions (phonological paraphasias)

- Target: "telephone" → Patient says: "telescope"
- Target: "butterfly" → Patient says: "buttercup"
- Target: "elephant" → Patient says: "elegant"

Sound-level errors (phonemic paraphasias)

- Target: "cat" → Patient says: "tat" or "gat" (consonant substitutions)
- Target: "spoon" → Patient says: "poon" or "s-poon" (consonant cluster reduction/struggle)
- Target: "banana" → Patient says: "manana" or "ba-nana" with effortful, groping articulation

Comprehension

Understanding of single words and simple sentences is usually relatively preserved. However, comprehension of complex or grammatically challenging sentences often breaks down, partly due to the underlying agrammatism.

Additional Features

- **Written Expression:** Writing mirrors spoken output, with reduced grammatical complexity and similarly telegraphic constructions.
- **Reading Aloud:** May reveal similar motor speech errors.
- **Social Function:** Patients may become frustrated or withdrawn due to reduced communicative ability, but insight is often relatively well maintained early on.
- **Other Cognition:** Memory, executive functioning, and visuospatial skills typically remain intact in the initial stages and only become affected as the disease progresses.

Differential Diagnosis in Nonfluent Agrammatic Primary Progressive Aphasia

Accurately differentiating nfaPPA from other conditions with overlapping symptoms is essential for the neuropsychologist. Key differentials include:

Semantic Variant PPA (svPPA)

- **svPPA** is marked by fluent speech with marked loss of word meaning and frequent semantic errors.
- Grammar and speech production are typically preserved, in contrast to the agrammatic, nonfluent speech of nfaPPA.

Logopenic Variant PPA (lvPPA)

- **lvPPA** is characterised by word-finding pauses, impaired repetition, and phonological errors, but without agrammatism or significant apraxia of speech.
- Grammar remains largely intact, and motor speech is fluent.

Post-Stroke Aphasias and Other Acquired Aphasia

- Sudden onset, rather than progressive, points to stroke or other acute causes.
- While Broca’s aphasia after stroke can resemble nfaPPA, neuroimaging and clinical history help distinguish degenerative from vascular causes.

Progressive Supranuclear Palsy and Corticobasal Syndrome

- Progressive supranuclear palsy and corticobasal syndrome can have overlapping motor speech or cognitive features. The presence of prominent parkinsonian features, gaze palsy, or apraxia beyond speech should raise suspicion for these syndromes.

Early Alzheimer’s Disease

- Alzheimer's Disease generally presents with early memory impairments, although language can become affected later.
- Insidious, isolated speech production and grammar disruption more strongly suggests nfaPPA.

Neuropsychological Assessment Considerations in Nonfluent Agrammatic Primary Progressive Aphasia

Clinical Interview

- Elicit the patient's typical communication patterns, difficulties in everyday contexts, and obtain a timeline of symptom progression.

Neuropsychological Language Testing

- **Expressive Language:**
  Assess for agrammatism (sentence construction, verb usage) and speech motor planning (praxis).
- **Motor Speech Examination:**
  Observe articulation, prosody, and sequence of sounds.
- **Repetition Tasks:**
  Note errors on multisyllabic words and phrases, and presence of apraxic errors.
- **Naming and Fluency Tasks:**
  While naming may be affected, expect fewer semantic errors compared to svPPA.
- **Comprehension Tests:**
  Gauge understanding of grammatically simple versus complex sentences.

Wider Cognitive Battery

- Screening for broader cognitive impairment, executive dysfunction, or changes in personality, to chart progression and support wider care planning.

Functional Impact

- Examine consequences for independence, activities of daily living, occupational function, and psychosocial wellbeing.

Neuroimaging and Pathology in Nonfluent Agrammatic Primary Progressive Aphasia

Structural and functional imaging often reveals asymmetric atrophy or hypometabolism in the left posterior frontal lobe (particularly the inferior frontal gyrus) and insular cortex. nfaPPA is usually

associated with **frontotemporal lobar degeneration**, often with **tau-positive pathology**, but can sometimes be linked to other underlying diseases.

Embedded media: https://radiopaedia.org/cases/24399/play?lang=gb

MRI Brain of a real nonfluent agrammatic primary progressive aphasia case in a 75-year old with "diagnosis probable" © Radiopaedia

Clinical Implications

Formulation and Feedback

- Provide a clear explanation of the nature of the disorder and likely progression.
- Support the patient and family to adapt communication methods, using simplified language, written cues, or alternative communication devices as progression occurs.

Intervention and Support

- Close liaison with speech and language therapists is critical.
- Psychoeducation, support groups, and practical planning for future care needs are important for this long-term, progressive condition.

Monitoring

- Regular neuropsychological and functional reviews enable timely adaptation of recommendations, assessment of decision-making capacity, and planning for emerging support needs.

Summary

Nonfluent/agrammatic primary progressive aphasia is a disorder of gradual onset and progression, defined by effortful, nonfluent speech and breakdown of grammar and sentence structure, often with associated apraxia of speech. For clinical neuropsychologists, careful assessment to differentiate nfaPPA from related conditions is essential, alongside ongoing support for patients, families, and care teams as the disease evolves.