# Frontotemporal dementia: a comparative case study of Greek-speaking individuals with the non-fluent and semantic variants of primary progressive aphasia

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## Introduction

Frontotemporal Dementia (FTD) is an umbrella term that encompasses degenerative disorders of the frontal and anterior temporal lobes that affect behavior and language. FTD overlaps clinically and pathologically with Primary Progressive Aphasia (PPA). PPA is a degenerative syndrome characterized by progressive loss of language function. The consensus criteria for PPA recognize three variants: the non-fluent/agrammatic variant of PPA (nfvPPA), the semantic variant of PPA (svPPA) and the logopenic variant of PPA (lvPPA) (Gorno-Tempini et al., 2011). Each PPA variant has a specific profile of language impairment, different distribution of atrophy on neuroimaging and different likelihood of underlying molecular pathology. Typically, nfvPPA is associated with fronto-insular atrophy, svPPA with atrophy of the anterior and inferior temporal lobe and lvPPA with atrophy of temporo-parietal regions. The most common types of neurodegeneration in PPA are frontotemporal lobar degeneration and Alzheimer's disease (Spinelli et al., 2017). FTD includes two out of three PPA variants, the nfvPPA and svPPA, as the most typical pathology of these variants is frontotemporal lobar degeneration.

PPA is characterized by a more partial and progressive pattern of damage than stroke-induced aphasia and targets areas such as the anterior temporal lobe that are rarely affected by stroke (Mesulam, 2016). Clinical and neuroimaging research on PPA has advanced our understanding of the language network. It has shown, for example, that the left anterior temporal lobe plays a critical role in single word comprehension and object naming and that the traditional 'Wernicke's area' is important for language repetition and sentence comprehension but not single word comprehension (Mesulam et al., 2019).

The aim of this study is to compare the clinical presentation of the language variants of FTD, nfvPPA and svPPA, in two Greek-speaking individuals with PPA. Greek is an underrepresented language in the literature on PPA research. It is a highly inflected and stem-based language. To this end, a comprehensive battery of neuropsychological tests and narrative analysis was employed.

## Methods

*Participants*

Two individuals diagnosed with PPA and 12 neurologically healthy adults participated in this study. All participants were right-handed.

The participant with the non-fluent variant of PPA, is a 61-year-old man with 6 years of formal education. He was assessed five years after symptom onset. He then had a sum of boxes score of 9 on the FTLD-modified Clinical Dementia Rating (CDR). MRI demonstrated left perisylvian atrophy.

The participant with the semantic variant of PPA is a 73-year-old man with 9 years of formal education. At the time of the study he had 5 years into the disease and a FTLD-modified CDR sum of boxes score of 6. His MRI scan showed the typical pattern of asymmetric anterior

temporal lobe atrophy.

The control group consisted of 2 male and 10 female native Greek speakers with a mean age of 68.08 (SD = 5.52) years and a mean of 13 (SD = 3.19) years of education.

*Procedure*
Participants were evaluated using a battery of tests assessing executive function, memory, visuospatial abilities, object semantics, mood, praxis, motor speech abilities, single word and sentence comprehension, repetition, confrontation naming, reading, writing and connected speech production. Quantitative production analysis (QPA) (Saffran et al., 1989) was used for the narrative analysis of a story retell task.

Neuropsychological testing was completed in four 45-minutes-sessions for the participants with PPA and three sessions for the control participants. MRI scans and reports were made available for the two individuals with PPA.

*Statistical analysis*
Crawford and Howell's method was used to compare performance of each subject with that of the control sample (Crawford, Garthwaite & Porter, 2010). T values were also calculated to compare the scores of the two subjects with reference to the control sample (Crawford, Garthwaite & Wood, 2010).

**Results**
The participant with the nfvPPA performed worse than the participant with the svPPA on the Digit Span –reverse recall task ($p$=0.025), Clock Drawing Test ($p$<0.001), syntactic comprehension (Boston Diagnostic Aphasia Examination, BDAE-3, $p$=0.014) and reading fluency for words ($p$<0.001).

The participant with the svPPA was more impaired in confrontation naming (Boston Naming Test-15, $p$<0.001), single word comprehension (Peabody Picture Vocabulary Test, $p$<0.001) and object semantics (Pyramid and Palm Trees Test, $p$=0.001). Comprehension of auditory complex material, written words and sentences were affected ($p$=0.022, $p$=0.005 and $p$<0.001, respectively), although his ability to follow commands was within normal limits and performance for syntactic comprehension was at ceiling. Both participants were impaired in the Trail Making Test A and B, verbal fluency, spelling and written picture description.

The narrative production measures that differed significantly between the two participants were speech rate (slower for the nfvPPA participant, $p$=0.007), average pause duration (longer for the nfvPPA participant, $p$<0.001), false starts per min (more for the nfvPPA participant, $p$=0.045), proportion of nouns (lower for the svPPA participant, $p$=0.012) and closed class words (lower for the nfvPPA participant, $p$=0.016). Compared to the control group, the nfvPPA participant produced shorter sentences ($p$=0.023), fewer closed class words ($p$=0.006), made longer pauses ($p$<0.001) and spoke at a slower rate ($p$<0.001). The svPPA participant used fewer nouns ($p$=0.027), more pronouns ($p$=0.02) and fewer narrative words as a proportion of the total words produced ($p$=0.003).

**Discussion**
The results confirm the distinctive features of both PPA variants, namely anomia, a single word comprehension deficit, preserved repetition and syntactic comprehension for the participant with the svPPA, as well as motor speech and syntactic processing difficulties alongside

with intact repetition, semantic knowledge and naming ability for the nfvPPA participant.

Taking into account the neuroimaging findings, these two cases illustrate the different distribution of atrophy in the language variants of FTD and highlight the role of the left anterior temporal lobe in naming and single word comprehension.

**References**

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