Definition of Primary Progressive Aphasia (PPA)

PPA is characterized by a progressive impairment of word usage and comprehension. Memory, personality, movement, face and object recognition remain relatively preserved for at least the first 2 years.

Core Features of PPA

- Naming deficits (anomia)
- Impaired fluency: word-finding pauses
- Agrammatism
- Impaired comprehension of word meaning

These features can be dissociated; any one is sufficient for diagnosis. PPA can be fluent or non-fluent.

The Core and Halo of PPA

Core Features (Language-Based)
- Impaired fluency: word-finding pauses
- Agrammatism
- Impaired comprehension of word meaning
- Naming deficits (anomia)

Boundary Features
- Dysarthria
- Idiomatic apraxia
- Dyscalculia
- Visual recognition deficits
- "Executive" dysfunction
- Poor memory for words—bu t event memory is preserved.
Longitudinal Investigations
In a PPA Patient Tested 6 and 9 years After Onset (M.K.)

Weintraub, Rubin & Mesulam, Archives of Neurology 1990.

Two MR Scans from the Same Patient Separated by a 3-yr Interval Mesulam, Annals of Neurology, 2001

73 yr Old Man With PPA

Mesulam, Annals of Neurology 2001

FUNCTIONAL MRI of LEXICAL PROCESSING
11 PPA VERSUS 11 AGE-MATCHED CONTROLS

Areas that were more activated in PPA than in controls during word identification tasks.

FLUENT PPA WITH POOR COMPREHENSION

MRI and 2-DG PET in Patient with PPA Onset at Age 42 (M. Sc.).

Voxel-Based Morphometry in Fluent vs Non-Fluent PPA

Non-fluent PPA

Fluent PPA Poor comprehension

Semantic Dysfunction in Frontotemporal Lobar Degeneration

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**Key Words**
Semantic dementia - Word meaning - Object recognition - Temporal lobes

**Abstract**
Semantic dementia, characterized by loss of word meaning and impaired object recognition, is one of the clinical manifestations of frontotemporal lobar degeneration and is associated with atrophy of the inferior and middle temporal gyri. Patients may present with problems in naming and understanding words or face and object recognition, the verbal and nonverbal nature of which reflects the anatomical location on the left or right temporal lobe. Behavioural changes

**Introduction**
In frontotemporal dementia (FTD), patients lose the capacity to interpret social content, instrumental functions, visual perception, and reality testing. The progression is less evident in general and eventually becomes evident. There is no evidence that they have difficulty in reading aloud or in recognizing other people. Some patients with FTD are in a severe condition in which they, despite the severity of the illness, still function in a profound loss of semantic. It may include loss of the ability to attend.

**NEUROPATHOLOGY OF PPA**

PPA belongs to the FTD-Pick-tauopathy spectrum of diseases, within which the major subtypes are PPA, SD and FLD.

**Possible association of the tau H1/H1 genotype With primary progressive aphasia**

**ApoE Allele Frequencies**

Medallion, Johnson, Grujic & Weintraub, *Neurology*, 1997


Krefft, Grotz-Okudara, Dickson, Baker, Castellani

One affected had DLDH, tau genotyping in another affected is negative.

Alberca, Montes, Russell, Grujic, Mesulam- Arch Neurol, in press

Left hemicanal craniosynostosis and hypoplasia. PPA onset at age 63.
PROBANDS AND FIRST DEGREE RELATIVES WITH LEARNING DISABILITY