Abstract
Primary progressive aphasia (PPA) is a clinical syndrome diagnosed when three core criteria are met. First, there should be a language impairment (i.e., aphasia) that interferes with the usage or comprehension of words. Second, the neurological work-up should determine that the disease is neurodegenerative, and therefore progressive. Third, the aphasia should arise in relative isolation, without equivalent deficits of comportment or episodic memory. The language impairment can be fluent or non-fluent and may or may not interfere with word comprehension. Memory for recent events is preserved although memory scores obtained in verbally mediated tests may be abnormal. Minor changes in personality and behavior may be present but are not the leading factors that bring the patient to medical attention or that limit daily living activities. This distinctive clinical pattern is most conspicuous in the initial stages of the disease, and reflects a relatively selective atrophy of the language network, usually located in the left hemisphere. There are different clinical variants of PPA, each with a characteristic pattern of atrophy. The underlying neuropathological diseases are heterogeneous and can include Alzheimer’s disease as well as frontotemporal lobar degeneration. The clinician’s task is to recognize PPA and differentiate it from other neurodegenerative phenotypes, use biomarkers to surmise the nature of the underlying neuropathology, and institute the most fitting multimodal interventions.

Keywords
dementia, language, network, frontotemporal, progranulin, tau.