

# Kyan Younes

## ***Right anterior temporal degeneration and loss of socioemotional semantics: The semantic behavioral variant frontotemporal dementia***

Focal anterior temporal lobe (ATL) degeneration often preferentially affects the left or right hemisphere. While patients with left-predominant ATL (lATL) atrophy show severe anomia and verbal semantic deficits and meet criteria for semantic variant primary progressive aphasia (svPPA), patients with early right ATL (rATL) atrophy are more difficult to diagnose. Uncertainty around early symptoms and absence of an overarching clinicoanatomical framework hinders proper diagnosis and care of patients with rATL disease.

We identified individuals from UCSF database with a clinical diagnosis of bvFTD or svPPA and a structural MRI (n=478). We defined three groups: rATL (n=46), frontal-predominant (n=79), and lATL-predominant (n=75). We compared the clinical, neuropsychological, genetic, and pathological profiles.

In the rATL-predominant group, the earliest symptoms were loss of empathy (27%), person-specific semantic impairment (23%), and complex compulsions and rigid thought process (18%). On testing, this group exhibited greater impairments in Emotional Theory of Mind, recognition of famous people, and facial affect naming. The early clinical symptoms were highly sensitive (81%) and specific (84%) differentiating rATL-predominant from frontal-predominant groups. FTLD-TDP (84%) was the most common pathology of the rATL-predominant group.

rATL-predominant degeneration is characterized by early loss of empathy and person-specific knowledge, deficits that are caused by progressive decline in semantic memory for concepts of socioemotional relevance. We outline new diagnostic criteria and propose the name, “semantic behavioral variant frontotemporal dementia” (sbvFTD). These diagnostic criteria will facilitate early identification and care of patients with early, focal rATL degeneration as well as in vivo prediction of FTLD-TDP pathology.

