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Right temporal variant frontotemporal dementia

There is growing consensus to consider the right temporal variant of frontotemporal dementia (rtvFTD) as a distinct type of FTD. Yet, lacking separate diagnostic criteria, patients with rtvFTD are often misdiagnosed with Alzheimer's disease or psychiatric disorders. Furthermore, based on neuroimaging features and non-verbal semantic impairment, it is also considered as a semantic variant primary progressive aphasia. Our previous work has shown that rtvFTD displayed a unique combination of cognitive (face recognition and memory impairments), behavioral/psychiatric (disinhibition, apathy, depression, hyper-religiosity, somatizations, and bizarre preoccupations), and language problems (word-finding difficulties and anomia). Moreover, preliminary findings in our cases and the literature review showed heterogeneity in the genetic and pathological background of rtvFTD. Since our single center data are not enough to elucidate the controversies on rtvFTD, with the contribution of FTD experts from 19 centers, we initiated an international multicenter study to shed line on the many unanswered questions about this equivocal syndrome. Our main goal is establishing international consensus criteria for the clinical diagnosis of rtvFTD based on the first and largest multinational rtvFTD cohort. Our sub-aims are elucidating the heritability, related genes/ underlying pathologies and the influence of amyloid pathology on the rtvFTD phenotype. We believe that the combination of our results and expert opinion will increase our knowledge about right temporal lobe neurodegeneration.

