Invited Address: Frontotemporal Dementias: Behaviour, Cognition, Phenotypes and Genotypes

Presenter: Julie Snowden

12:00–1:00 p.m.

J. SNOWDEN. Frontotemporal Dementias: Behaviour, Cognition, Phenotypes and Genotypes.

The frontotemporal dementias are clinically and pathologically heterogeneous. The predominant symptoms may be of problems in behavior, executive skills, language or conceptual knowledge. A proportion of patients show physical signs of amyotrophic lateral sclerosis. It is well recognized that patients' behavioural/cognitive profile reflects the anatomical distribution of degenerative change within the anterior hemispheres. There is, however, growing evidence that it is also influenced by the type of underlying pathology and by genetic mutations associated with frontotemporal dementia. The talk considers the neuropsychological variation within the frontotemporal dementias and examines its relationship to pathology and genetics. Systematic associations are demonstrated, which suggest that behavioural/cognitive profiles, taken together with other clinical features, are predictors of pathology and genetic status. The talk shows the importance of neuropsychology in delineating the diversity of clinical phenotypes in frontotemporal dementia. It is argued that neuropsychology has a crucial role not only in clinical diagnosis and management of patients with frontotemporal dementia, but also in the theoretical understanding of disease mechanisms.

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