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Current Position

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Keywords

Neuropathology – Frontotemporal Lobar Degeneration – Amyotrophic Lateral Sclerosis - proteinopathies

Science

Frontotemporal lobar degeneration (FTLD) is the second most common form of young onset dementia, but it has also a high prevalence in the elderly population. As our population is getting older, getting a better understanding of this disease, within its spectrum of neurodegenerative disorders, is mandatory. We build further on very important research data, which elicited the presence of TDP-43 and FUS inclusions, thus causing FTLD and the related motor neuron disorders (amyotrophic lateral sclerosis - ALS). At this moment, different gene mutations have been described, causing typical morphology and localization of the inclusions (C9orf72, GRN, VCP). However, there remains a group of FTLD patients which do have ubiquitin immunoreactive inclusions, which do not stain positive for TDP-43 or FUS. These inclusions stain positive with p62 and ubiquilin. This could elicit additional information which may be specific for gene mutations. Further, co-localisation of other inclusions, suggestive for other neurodegenerative disorders will be searched for in combination with the TDP-43 and FUS lesions. Our extensive sampling and immunohistochemical examination of the brain and spinal cord of patients with FTLD and/or ALS will have add an extra value for the data now published.

Recent Research Projects

Research Foundation Flanders (FWO) - Clinical Grant
Period: 15.11.2010 – 01.02.2013
Role:

Selected Publications

Gijselinck I, Van Langenhove T, van der Zee J, Sleegers K, Philtjens S, Kleinberger G, Janssens J, Bettens K, Van Cauwenberghe C, Pereson S, Engelborghs S, **Sieben A**, De Jonghe P, Vandenberghe R, Santens P, De Bleecker J, Maes G, Bäumer V, Dillen L, Joris G, Cuijt I, Corsmit E, Elinck E, Van Dongen J,

Vermeulen S, Van den Broeck M, Vaerenberg C, Mattheijssens M, Peeters K, Robberecht W, Cras P, Martin JJ, De Deyn PP, Cruts M, Van Broeckhoven C. A C9orf72 promoter repeat expansion in a Flanders-Belgian cohort with disorders of the frontotemporal lobar degeneration-amyotrophic lateral sclerosis spectrum: a gene identification study. Lancet Neurol. 2012 Jan;11(1):54-65. (PMID: 22154785)

Sieben A, Van Langenhove T, Engelborghs S, Martin JJ, Boon P, Cras P, De Deyn PP, Santens P, Van Broeckhoven C, Cruts M.The genetics and neuropathology of frontotemporal lobar degeneration. Acta Neuropathol. 2012 Sep;124(3):353-72. Epub 2012 Aug 14. (PMID: 22890575)

All Publications