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IN BRIEF

MULTIPLE SCLEROSIS

IgM bands indicate reduced risk of PML on natalizumab

A new study has linked the presence of IgM oligoclonal bands in the cerebrospinal fluid with decreased risk of progressive multifocal leukoencephalopathy (PML) during natalizumab treatment for multiple sclerosis. The researchers report the risk reduction to be independent of anti-JC-virus antibody status. PML is a serious adverse effect associated with natalizumab treatment; in most patients, anti-JC antibodies increase the risk of PML.

Original article Villar, L. M. *et al.* Lipid-specific IgM bands in CSF associated with a reduced risk of developing PML during treatment with natalizumab. *Ann. Neurol.* doi:10.1002/ana.2434

NEURODEGENERATIVE DISEASE

Acetylation controls TDP-43 function and aggregation

Post-translational acetylation of TDP-43 promotes accumulation of insoluble, hyperphosphorylated TDP-43, according to a new study. Accumulation and propagation of TDP-43 pathology is a hallmark sign of amyotrophic lateral sclerosis (ALS) and frontotemporal lobar degeneration (FTLD). Modulation of TDP-43 acetylation could, therefore, provide a strategy to intervene in the pathogenesis of these disorders.

Original article Cohen, T. J. *et al.* An acetylation switch controls TDP-43 function and aggregation propensity. *Nat. Commun.* doi:10.1038/ncomms6845

SENSORY SYSTEMS

Cochlear implantation can ameliorate symptoms of OPA1-associated auditory neuropathy

OPA1 is best known for its association with optic atrophy, but it can also cause auditory neuropathy. In a new study, Santarelli and colleagues report impaired speech perception and abnormal auditory brainstem responses in patients with OPA1 missense mutations. The researchers posit that OPA1-associated auditory neuropathy is caused by neural degeneration that affects the terminal dendrites and disrupts synchrony in the auditory nerve fibre. Cochlear implants, which bypass the site of lesion, improved auditory function in most of these patients.

Original article Santarelli, R. *et al.* OPA1-related auditory neuropathy: site of lesion and outcome of cochlear implantation. *Brain* doi:10.1093/brain/awu378

DEMENTIA

Improved decision tree aids differentiation of atypical Alzheimer disease from Creutzfeldt–Jakob disease

Atypical phenotypes of Alzheimer disease (AD) can sometimes be difficult to distinguish from Creutzfeldt–Jakob disease (CJD). Dorey and colleagues report that the level of total prion protein (t-PrP) in the cerebrospinal fluid (CSF) is lower in patients with CJD than in patients with atypical AD. In differential diagnosis of CJD and atypical AD, CSF t-PrP yielded 82.1% sensitivity and 91.3% specificity. Dorey *et al.* suggest a diagnostic decision tree that combines analysis of CSF t-PrP with other nonspecific biomarkers, which could decrease the misclassification rate of atypical AD phenotypes to as low as 4.3%.

Original article Dorey, A. *et al.* Association of cerebrospinal fluid prion protein levels and the distinction between Alzheimer disease and Creutzfeldt–Jakob disease. *JAMA Neurol.* doi:10.1001/jamaneurol.2014.4068