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IN BRIEF

WHITE MATTER DISEASE

Hyaluronidase PH-20 digestion products inhibit remyelination in demyelinating lesions

The products of hyaluronan digestion by hyaluronidase PH-20 inhibit oligodendrocyte progenitor cell (OPC) maturation in demyelinating lesions, leading to failure of remyelination, new research shows. PH-20 was found to be upregulated in demyelinating lesions in both mice and humans, and inhibition of this enzyme promoted OPC maturation in cell culture and in the mouse brain. The authors propose that PH-20 could be a therapeutic target in demyelinating diseases such as multiple sclerosis.

Original article Preston, M. *et al.* Digestion products of the PH20 hyaluronidase inhibit remyelination. *Ann. Neurol.* doi:10.1002/ana.23788

EPILEPSY

EEG findings correlate with hippocampal injury in children with febrile status epilepticus

Focal slowing or attenuation of EEG activity within 72 h of febrile status epilepticus (FSE) in children could be indicative of acute hippocampal injury, according to the latest findings of the FEBSTAT study. Of 199 children assessed within 72 h of FSE presentation, 45.2% exhibited abnormal EEG activity. Focal slowing and attenuation were found to be strongly associated with hippocampal T2 signal abnormalities on MRI.

Original article Nordli, D. R. Jr *et al.* Acute EEG findings in children with febrile status epilepticus: results of the FEBSTAT study. *Neurology* doi:10.1212/WNL.0b013e3182759766

MOTOR NEURON DISEASE

N-type Ca^{2+} channels are implicated in amyotrophic lateral sclerosis

Research from Italy indicates that N-type Ca^{2+} channels contribute to the pathogenesis of amyotrophic lateral sclerosis (ALS). These channels were found to be overexpressed in cortical neurons and motor cortical tissue from mice with the ALS-related mutation Gly93Ala in the *SOD1* gene. Compared with control cells, Gly93Ala cortical neurons showed increased calcium entry during depolarization, as well as reduced survival—an effect that was reversed by addition of an N-type Ca^{2+} channel blocker.

Original article Pieri, M. *et al.* Over-expression of N-type calcium channels in cortical neurons from a mouse model of amyotrophic lateral sclerosis. *Exp. Neurol.* doi:10.1016/j.expneurol.2012.11.002

NEUROMETABOLIC DISEASE

Proteomic analysis identifies biomarkers for Niemann–Pick disease, type C1

A quantitative proteomic study has revealed dysregulation of a number of proteins in Niemann–Pick disease, type C1 (NPC1). An initial analysis in mouse cerebellar tissue identified 77 proteins that were differentially expressed between *Npc1* mutants and controls. Altered expression of three of these proteins—glutathione *S*-transferase α, superoxide dismutase and FABP3—was confirmed in cerebrospinal fluid from patients with NPC1.

Original article Cologna, S. M. *et al.* Quantitative proteomic analysis of Niemann–Pick disease, type C1 cerebellum identifies protein biomarkers and provides pathological insight. *PLoS ONE* **7**, e47845 (2012)