



In Focus

Spotlight on the November 18 Issue

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TLR3 deficiency in herpes simplex encephalitis: High allelic heterogeneity and recurrence risk

By identifying the novel *TLR3* mutations in 120 children with herpes simplex encephalitis (HSE), the authors determined the proportion of HSE patients (5%) with a mutation in *TLR3*. HSE of childhood is not purely a virally determined illness but may result from genetic determinism of inborn errors of *TLR3* impairing CNS intrinsic immunity to herpes simplex virus-1.

See p. 1888

From editorialists Steiner & Tyler: "The observation that other genes associated with innate immunity were defective in some patients with HSE having normal *TLR3*... suggests that inborn innate immunity defects at large predispose to HSE."

See p. 1882

Both aerobic exercise and cognitive-behavioral therapy reduce chronic fatigue in FSHD: An RCT ▲

Forty-two aerobic exercise training (AET) and 5 cognitive behavior therapy (CBT) sessions were more effective than usual care for reducing fatigue and improving physical activity in patients with facioscapulohumeral muscular dystrophy. Both AET and CBT may be incorporated in a treatment program for reducing fatigue in these patients.

See p. 1914

Protective environmental factors for neuromyelitis optica

The authors examined early-life exposures to common herpes viruses in 36 pediatric patients with neuromyelitis optica (NMO), 491 with multiple sclerosis (MS), and 224 controls. Exposure to other young children was consistent with the hypothesis that infections contribute to disease risk reduction, but unlike MS, pediatric NMO is not associated with exposures to common herpes viruses.

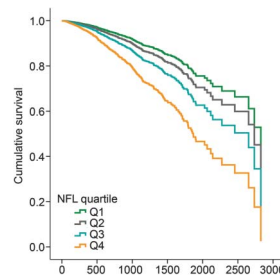
See p. 1923

Increased CSF APPs- α levels in patients with Alzheimer disease treated with acitretin ▲

Promoting nonamyloidogenic amyloid precursor protein processing via α -secretase activation could open new therapeutic avenues for Alzheimer disease. Treatment of Alzheimer disease patients with acitretin (30 mg daily) for 4 weeks increased α -secretase activity and was well-tolerated. Acitretin is a promising new therapeutic option for Alzheimer disease; however, determination of clinical efficacy is needed.

See p. 1930

CSF neurofilament light differs in neurodegenerative diseases and predicts severity and survival



This study analyzed CSF levels of neurofilament light in relation to disease severity, survival, and other biomarker levels in 3,356 patients with dementia. High levels of neurofilament light were associated with more severe disease and shorter survival, and levels were

especially high in disorders affecting subcortical regions of the brain.

See p. 1945

Determinants of antithrombotic choice for patent foramen ovale in cryptogenic stroke

The authors examined 1,132 patients with cryptogenic stroke (CS) and patent foramen ovale (PFO) treated with anticoagulation or antiplatelet agents. Both antithrombotic regimens were used for secondary stroke prevention in patients with CS and PFO. Radiologic and echocardiographic features were associated with treatment choice whereas conventional vascular risk factors were not.

See p. 1954

VIEWS & REVIEWS

Dietary treatment in adults with refractory epilepsy:

A review 📖 📖

There is growing interest in treatment of refractory epilepsy with ketogenic-based diets in adults. In the small studies reviewed here, the diets were well-tolerated, and approximately 30% of patients achieved $\geq 50\%$ seizure reduction and 5%-10% achieved $\geq 90\%$ seizure reduction; however, social and culinary restrictiveness limited their acceptance and adherence. Larger, controlled studies are needed.

See p. 1978

NB: "Apnea and dysphagia as the sole features of an α -synucleinopathy," see p. 1988. To check out other Clinical/Scientific Notes, point your browser to Neurology.org.

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