

In Focus

Spotlight on the July 6 Issue

Robert A. Gross, MD, PhD, FAAN
Editor-in-Chief, *Neurology*[®]

High insulinlike growth factor I is associated with cognitive decline in Huntington disease

This study determined fasting blood levels of total insulinlike growth factor I (IGF1), growth hormone, and insulinlike factor binding protein 3 at baseline in 109 patients with genetically documented Huntington disease. The findings suggest that a high total IGF1 level at baseline may be associated with greater subsequent declines in executive function and attention.

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Clinical and biochemical features of aromatic L-amino acid decarboxylase deficiency



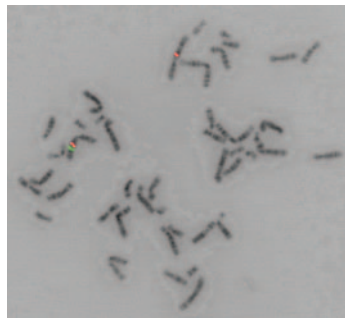
The authors assessed the clinical and biochemical data of 78 patients with aromatic L-amino acid decarboxylase deficiency from a database of pediatric neurotransmitter disorders. Only 15 patients with a relatively mild form improved on a combined therapy with pyridoxine (B6)/pyridoxal phosphate, dopamine agonists, and MAO-B inhibitors.

See p. 64; Editorial, p. 15

Four generations of epilepsy caused by an inherited microdeletion of the SCN1A gene



The authors performed a clinical interview, neurologic examination, and electrophysiologic testing in a 4-generation Bulgarian family with 4 individuals with generalized epilepsy. They found that not only patients with severe myoclonic epilepsy of infancy (SMEI), but also patients with inherited severe generalized



epilepsy with febrile seizures plus (GEFS+), need to be screened for copy number variations in SCN1A.

See p. 72; Editorial, p. 18

VISIONS

In this issue we initiate a new feature, Visions, that will showcase artistic visual works on medical and scientific themes. The first offering is titled "Three Flasks."

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VIEWS & REVIEWS

The case against confirmatory tests for determining brain death in adults



A confirmatory test is not mandatory, but is useful as a safeguard, added when findings on clinical examination are incomplete, in determining brain death in adults. This review discusses the origin of confirmatory tests, the accuracy of the tests, and the consequences of not using them.

See p. 77

CONTEMPORARY ISSUES IN NEUROLOGIC PRACTICE

Psychogenic nonepileptic "seizures" or "attacks"? It's not just semantics

The first step in treatment is diagnosis. These articles discuss the importance of the words used in conveying the diagnosis of psychogenic nonepileptic events and why we should do away with the confusing word "seizure" when we refer to psychogenic episodes.

See pp. 84 and 87

CLINICAL/SCIENTIFIC NOTES

De novo relapsing-remitting multiple sclerosis following autologous stem cell transplantation

The authors examined a 62-year-old man with no pre-BMT MRI abnormality who developed relapsing-remitting multiple sclerosis. These observations may be pertinent to the role of immune dysregulation in multiple sclerosis and the consequences of immune dysregulation following induced lymphopenia or BMT, undertaken for this very condition.

See p. 89

AAN AFTER-DINNER PRESENTATION BY DR. ROBERT JOYNT

Dr. Robert Joynt was interviewed by Dr. Ted Burns, Podcast Section Editor, about his presentation at the AAN Leadership Dinner. Dr. Joynt recounts his personal relationship with the founders of the AAN. Please check out this special interview that will be posted as part of the July 6, 2010, Podcast Session at <http://www.aan.com/rss/?event=feed&channel=1>. AAN members can obtain free CME credits after listening to this humorous history lesson (go to www.neurology.org).

Podcasts can be accessed at www.neurology.org

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