August 14 Highlights

Coenzyme Q₁₀ (CoQ₁₀) in Huntington’s disease and myopathy

“No significant benefit... but a tantalizing trend favoring CoQ₁₀”

Two papers and an editorial consider the use of CoQ₁₀. The Huntington Study Group’s 30-month, randomized, controlled trial of CoQ₁₀ and remacemide for HD found no significant benefit of either or both drugs for the primary end point, total functional capacity. There was a tantalizing trend (p = 0.15) favoring CoQ₁₀ use. The Di Giovanni et al. study of familial CoQ₁₀ deficiency showed dramatic benefit of CoQ₁₀ on symptoms and biopsy—resolution of lipid storage, increased mitochondria and mitochondrial enzymes, and abrogation of apoptotic histology.

In the accompanying editorial, Shults and Shapira note that CoQ₁₀ is clearly a specific treatment for the rare patients with its deficiency. Whether CoQ₁₀ has a broader role in degenerative disease is not settled and merits additional study.

Deficient catechol-O-methyltransferase (COMT) and velo-cardio-facial syndrome

One copy of the COMT gene is absent in the 22q11.2 deletion syndrome. If the nondeleted chromosome encodes a variant of COMT with low stability, increased catecholamine neurotransmission could contribute to the neuropsychiatric features, which include a severe bipolar disorder. Graf et al. describe patients with the deletion syndrome in whom CSF catecholamine levels were measured and report that lowering catecholamine production with metyrosine corrects the defect and may alleviate symptoms—an observation that should prompt a controlled treatment trial.

In the accompanying editorial, Percy and Nemeroff note that because a severe bipolar disorder is part of the chromosome 22q11.2 deletion syndrome, molecular genetics and psychopharmacology can now bridge the mind–brain interface, providing insights into causes and treatments for psychiatric disorders.

Sudden unexplained death in epilepsy (SUDEP) in children

Donner et al. report a large series of SUDEP in children. Previously established risk factors for SUDEP in adults, including low serum antiepileptic drug levels and antiepileptic drug polytherapy, were not significant risk factors for SUDEP in children.

Epilepsy: A major traffic risk?

Lings et al. compared Danish subjects with epilepsy with controls individually matched for age, sex, place of residence, and exposure period. Patients with epilepsy were seven times more likely to have been treated at a casualty department after a driving accident.

Donepezil in progressive supranuclear palsy (PSP): Minimal benefit and possible worsening

The Litvan et al. randomized, double-blind, crossover trial compared donepezil with placebo in 21 patients with PSP. Memory scores did not improve significantly, and mobility scores worsened. Donepezil is not recommended for this patient population.
**Constipation: Risk factor or result of Parkinson’s disease?**

“Further study is needed to determine whether constipation is part of early PD or a marker of factors that cause PD.”

Abbott et al. examined the relation between bowel movement frequency and risk of subsequently developing Parkinson’s disease in 6790 asymptomatic men aged 51 to 75 years prospectively studied in the Honolulu Heart Program. After 24 years of follow-up, infrequent bowel movements were associated with a threefold elevated risk of Parkinson’s disease in later life.

*see page 456*

**Donepezil in AD: Effects on function during a year of treatment**

Patients with AD gradually lose independent functioning of basic and instrumental activities of daily living. In a parallel group, double-blind study, Moohs et al. found that patients with AD treated with donepezil were 38% less likely to suffer a clinically significant loss of function during 1 year than were patients treated with placebo. Median time to a significant loss of function was delayed by 5 months in the patients treated with donepezil.

*see page 481*

**Long-term benefit of donepezil in mild to moderate AD**

The Winblad et al. 1-year multinational, parallel group, placebo-controlled study of donepezil in patients with AD found the drug well tolerated and effective in terms of global symptoms of dementia, activities of daily living, and cognition; cognition was maintained close to baseline in patients treated with donepezil for 1 year.

*see page 489*

**Lamotrigine for painful diabetic neuropathy (PDN)**

PDN is often refractory to treatment. Eisenberg et al. conducted a parallel group study of lamotrigine in 60 patients with PDN. They found that lamotrigine was efficacious in PDN, confirming results of phase II trials in PDN and phase III trials of lamotrigine for other painful neuropathies.

*see page 505*

**Delirium in the ICU from nicotine withdrawal**

Mayer et al. describe five neuro-ICU patients with agitated delirium, all heavy cigarette smokers, who experienced immediate and dramatic improvement of their symptoms after application of a 21-mg nicotine patch.

*see page 551*

**Prospagnosia from right temporal lobe disease with olfactory agnosia**

*Agnosia for familiar faces and odors in a patient with right temporal lobe dysfunction*

Mendez and Ghajarnia studied a patient with both progressive prosopagnosia and inability to identify odors. Results of their study suggest that the right temporal lobe is needed for recognition of odors and faces.

*see page 519*

*The professor or the resident?*

The repeated inability of a patient with a right temporo-occipital stroke to distinguish just two faces prompted this report by Nyffeler et al. The patient did better if shown the two faces inverted. The puzzling selectivity of the defect prompts consideration of paramnesic misidentification, Capgras and Fregoli syndrome, and intermetamorphosis.

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