

In 1972, Livingston summarized the literature on the effectiveness of d-amphetamine in the control of “petit mal” and other epilepsies. For children <6 years old, Livingston recommended initial d-amphetamine doses of 2.5 mg daily, and >6 years, 2.5 mg 2 x daily (Livingston S. **Comprehensive Management of Epilepsy in Infancy, Childhood and Adolescence**. Springfield, IL, Charles C Thomas, 1972, pp. 198 and 298-300). Given the adverse publicity associated with fenfluramine, future trials of stimulants in Dravet syndrome patients might substitute d-amphetamine (l-amphetamine is found ineffective as an anticonvulsant). For the early recognition and when to suspect the diagnosis of Dravet syndrome, see a review from the Comprehensive Epilepsy Center, Ann & Robert H. Lurie Children’s Hospital of Chicago (Millichap JJ, Koh S, Laux LC, Nordli DR Jr. **Neurology** 2009 Sep 29;73(13):e59-62).

## **METABOLIC DISORDERS**

### **GABA-ERGIC DYSFUNCTION IN SUCCINIC SEMIALDEHYDE DEHYDROGENASE DEFICIENCY**

Investigators at the Clinical Epilepsy and Neurorehabilitation Sections, NIH, Bethesda, MD; Albert Ludwigs University, Freiberg, Germany; and Children’s National Medical Center, Washington, DC used transcranial magnetic stimulation (TMS) to quantify the excitation and inhibition in primary motor cortex in 8 patients (mean age 15.4 years) with succinic semialdehyde dehydrogenase (SSADH) deficiency. All patients were severely affected and many showed symptoms of ADHD and anxiety. Long interval intracortical inhibition was significantly reduced and the cortical silent period was significantly shortened in patients with SSADH deficiency compared to heterozygous parents and controls. Long interval intracortical inhibition and cortical silent period are thought to reflect GABA receptor-mediated inhibitory circuits, pointing to a GABA-ergic motor cortex dysfunction in patients with SSADH deficiency. (Reis J, Cohen LG, Pearl PL, et al. GABAB-ergic motor cortex dysfunction in SSADH deficiency. **Neurology** 2012 Jul 3;79(1):47-54). (Response: Dr Reis. E-mail: janine.reis@uniklinik-freiburg.de).

COMMENT. SSADH deficiency is a rare autosomal recessive disorder of GABA degradation with elevation of gamma-hydroxybutyric acid and GABA. Infants present with developmental delay, hypotonia, retardation, ataxia, seizures, hyperkinetic behavior, aggression, and sleep disturbances. Urine organic acids show 4-hydroxybutyric/gamma-hydroxybutyric aciduria. MRI may show globus pallidus T2 abnormalities. TMS may be helpful in detection of homozygous carriers and in diagnosis of SSADH deficiency.

## **INFECTIOUS DISORDERS**

### **RESIDENTS’ LUMBAR PUNCTURE SKILLS AFTER SIMULATION-BASED EDUCATION**

Researchers in the Departments of Medicine and Neurology at Northwestern University Feinberg School of Medicine, Chicago, IL evaluated the effect of simulation-

based mastery learning (SBML) on internal medicine residents' lumbar puncture (LP) skills, assessed neurology residents' acquired LP skills from traditional clinical education, and compared the results of SBML to traditional clinical education. Residents completed a baseline skill assessment (pretest) and following a 3-hour practice and feedback, residents completed a posttest. PGY1 internal medicine residents (n=58) improved from a mean of 46.3% to 95.7% after SBML (p<001) and all met the minimum passing score (set by an expert panel) at final posttest. PGY2, 3 and 4 neurology residents (n=36) from 3 medical centers completed the same simulated LP assessment without SBML. The performance of traditionally trained neurology residents was significantly lower than simulator-trained residents (mean 65.4%, p<0.001) and only 6% met the minimum passing score. (Barsuk JH, Cohen ER, Caprio T, McGaghie WC, Simuni T, Wayne DB. Simulation-based education with mastery learning improves residents' lumbar puncture skills. *Neurology* 2012 Jul 10;79(2):132-7). (Response & reprints: Dr Barsuk. E-mail: jbarsuk@nmh.org).

COMMENT. Internal medicine residents who complete SBML show improvement in LP skills, with competence superior to that of traditionally trained neurology residents. Simulation technology increases procedural skill by providing practice opportunity in a relaxed environment. The researchers recommend that residents complete simulation-based training prior to performing a clinical LP. The medical maxim that experience treating the patient translates to procedural expertise is disproven for competence in performing LP (Nathan BR, Kincaid O. Does experience doing lumbar punctures result in expertise? A medical maxim bites the dust. *Neurology* 2012 Jul 10;79(2):115-6). Simulator training and adherence to a procedural checklist are important in the preparation and performance of an LP by residents.

## **RISK OF GUILLAIN-BARRE SYNDROME AND H1N1 INFLUENZA VACCINATION**

Researchers at Laval University, Department of Pediatric Neurology (Dr Boucher), Quebec University Hospital and other centers in Montreal, Quebec, Canada assessed the risk of Guillain-Barre syndrome (GBS) following the pandemic influenza vaccination campaign in 2009, mostly using an AS03 adjuvant vaccine. Over a 6-month period, 83 confirmed GBS cases were identified and 25 had been vaccinated against 2009 influenza A (H1N1) 8 or fewer weeks before disease onset (most (19/25) were vaccinated 4 or fewer weeks before onset). Relative risk was 1.80 for all cases during the 8-week postvaccination period and 2.75 during the 4-weeks postvaccination. The number of GBS cases attributable to vaccination was 2 per 1 million doses. No confirmed case of GBS was reported in the immunized age group 6mo-9years; one case occurred in the 10-19 year old immunized group (rate 1.20 per 100,000 person-years). (De Wals P, Deceuninck G, Toth E, et al. Risk of Guillain-Barre syndrome following H1N1 influenza vaccination in Quebec. *JAMA* 2012 Jul 11;308(2):175-81). (Respond: Philippe De Wals MD PhD, Universite Laval, Quebec. E-mail: philippe.de.wals@sss.gov.qc.ca).

COMMENT. A small but significant risk of GBS was associated with the 2009 influenza A (H1N1) vaccination campaign in Quebec, Canada. In the US, results of the