

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. GABA-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-