

SEIZURE DISORDERS

GLUCOSE TRANSPORTER 1 DEFICIENCY AND MYOCLONIC ASTATIC EPILEPSY

Researchers at University of Melbourne, Australia; University of Florence, Italy; and University of Antwerp, Belgium performed genetic analysis on 84 unrelated probands with myoclonic-astatic epilepsy (MAE). Seizures began at 8 – 48 months of age. EEG showed generalized spike and wave. Cognitive impairment was progressive, except in one patient who received the ketogenic diet early. Four patients had a mutation of *SLC2A1* on sequencing. Two patients with MAE and *SLC2A1* mutations also developed paroxysmal exertional dyskinesia in childhood. (Mullen SA, Marini C, Suls A, et al. Glucose transporter 1 deficiency as a treatable cause of myoclonic astatic epilepsy. *Arch Neurol* 2011;68(9):1152-1155). (Respond: Renzo Guerrini MD, Children's Hospital A Meyer-University of Florence, Viale Pierraccini 24, 50139 Firenze, Italy. E-mail: r.guerrini@meyer.it).

COMMENT. Sequencing of *SLC2A1* as part of the workup for MAE is expected to uncover 5% patients who test positive and should be responsive to the ketogenic diet.

LATE ONSET ICTAL ASYSTOLE IN REFRACTORY EPILEPSY

Researchers at Albert Einstein College of Medicine, Bronx, NY report a 17-year-old boy with chronic intractable epilepsy and vagus nerve stimulator who developed ictal asystole many years after the onset of epilepsy. The boy had severe mental retardation, autism, and refractory epilepsy throughout childhood. MRI showed symmetric posterior cortical polymicrogyria. A first episode of supraventricular tachycardia occurred at 11 years old, and a vagus nerve stimulator inserted at age 12 years. On long-term video-EEG monitoring and cardiac telemetry, tonic arm extension seizures with head deviation to either side were accompanied by a 3-9 second period of asystole, simultaneous with attenuation of the EEG. The EEG indicated frequent electrographic seizures confined to the right posterior hemisphere quadrant. Persistent baseline bradycardia 50-65 beats per min and occasional sinus pauses lasting 1-2 secs were not associated with clinical or EEG seizures. The asystole was not linked to the vagus nerve stimulator, and ultimately necessitated placement of a cardiac pacemaker, with no interaction of the two devices. Repeated seizures may injure the brain parasympathetic or cardiac centers, leading to asystole. (Beal JC, Sogawa Y, Ceresnak SR, Mahgerefteh J, Moshe SL. Late onset ictal asystole in refractory epilepsy. *Pediatr Neurol* Oct 2011;45:253-255). (Respond: Dr Beal, Department of Child Neurology, Montefiore Medical Center, 111 East 210th Street, Bronx, NY 10467. E-mail: jbeal@montefiore.org).

COMMENT. Cardiac abnormalities associated with epilepsy include tachycardia or bradycardia, T-wave flattening, AV block, atrial fibrillation, long QT, torsades de