

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

pointe (paroxysmal ventricular tachycardia), and ictal asystole. Asystole has a role in sudden unexplained death in epilepsy.

Ictal asystole and anti-NMDAR antibody encephalitis. Ictal asystole is recently reported as a complication of anti-NMDAR encephalitis. In this 15-year-old girl, seizures with asystole developed 26 days after initial presentation of symptoms and temporal lobe seizures that were associated with bradycardia. After insertion of a demand pacemaker on day 46, there were no further cardiac events. (Millichap JJ, Goldstein JL, Laux LC, Nordli DR, Stack CV, Wainwright MS. *Pediatrics* 2011;127(3):e781-6). In this case, asystole occurred at the onset of the illness and was not explained by a prolonged recurrence of seizures.

FUNCTIONAL NEUROIMAGING IN STARTLE EPILEPSY

Researchers at the Epilepsy Unit, Hospital Clinic de Barcelona, Spain investigated brain areas involved in startle-induced seizures, using a functional neuroimaging approach in 4 adult patients whose seizures began at age 4 months to 10 years. Presurgical evaluation included ictal SPECT coregistered to MRI. Startle-induced seizures were bilateral asymmetric tonic with ictal-EEG pattern located over the mesial centroparietal region. Three patients had a significant hyperperfusion (> 2 SD above the reference) involving the supplementary motor area, the perirolandic area, and precuneus. Ictal EEG-fMRI showed an initial activation located over the precuneus, supplementary motor area, cingulate gyrus, and the precentral/perirolandic area. Startle-induced seizures triggered by unexpected stimuli are generated by the interaction of a frontoparietal network located over the mesial brain surface. (Fernandez S, Donaire A, Maestro I, et al. Functional neuroimaging in startle epilepsy: Involvement of a mesial frontoparietal network. *Epilepsia* Sept 2011;52(9):1725-1732). (Respond: Santiago Fernandez MD, E-mail: santiago.fernandez@hospitalplato.com).

COMMENT. Startle epilepsy is triggered by unexpected stimuli, generally a sudden noise, somatosensory, or visual stimuli. First described by Alajouanine and Gastaut (1955), and included in the ILAE classification 2001. A frontoparietal epileptic network is involved, and not a discrete focus.

ATTENTION DEFICIT COMORBID DISORDERS

WRITTEN-LANGUAGE DISORDER AND ADHD

Researchers at Mayo Clinic, Rochester, MN, and Children's Hospital Boston, MA evaluated the incidence rates of written-language disorder (WLD), with and without reading disability (RD), according to gender, among children with and without attention deficit hyperactivity disorder (ADHD) in a population-based birth cohort. The cumulative incidence of WLD by 19 years of age was significantly higher for children with ADHD than for children without ADHD, for both boys and girls (boys: 64.5% vs 16.5%; girls: 57.0% vs 9.4%). In contrast, among children without ADHD, the