

MRI and 1H-MRS may prove effective in assessment of effects of cholesterol replacement therapy in patients with SLOS (Caruso PA et al. **Neuroradiology** 2004 Jan;46(1):3-14). Of 18 patients with SLOS, abnormal CNS findings were noted in 5 patients, including callosal abnormalities in 4 (22%), Dandy-Walker variant in 1, arachnoid cyst in 1, and holoprosencephaly in 1 (6%). Clinical degree of disease severity was correlated with lipid:choline ratios.

DEVELOPMENTAL OUTCOME FOLLOWING HEMISPHERECTOMY FOR HEMIMEGALENCEPHALY

Investigators at the National Center of Neurology and Psychiatry, Tokyo, studied the effect of early hemispherectomy on development in a consecutive series of 12 infants with hemimegalencephaly (HME) and epileptic encephalopathy. Mean age at onset was 20.4 days, mean age at surgery was 4.3 months (range 2-9), and mean follow-up time was 78.8 months (36-121). Eleven patients had a history of early infantile epileptic encephalopathy. Following vertical parasagittal hemispherectomy, 8 (66.7%) were seizure-free and showed significantly higher postoperative developmental quotient (DQ) than those with seizures (mean 31.3 vs 5.5; $p=0.02$). In the seizure-free group, postoperative DQ correlated with postoperative seizure duration ($p=0.01$). Shorter seizure duration during early infancy provides better postoperative DQ in infants with HME and epileptic encephalopathy. (Honda R, Kaido T, Sugai K, et al. Long-term developmental outcome after early hemispherectomy for hemimegalencephaly in infants with epileptic encephalopathy. **Epilepsy Behav** 2013 Aug 6;29(1):30-35). (Response: Dr Kaido. E-mail: kaido@ncnp.go.jp).

COMMENT. Despite the risks, the authors conclude that early surgical intervention for hemimegalencephaly is preferable to brain damage from repeated seizures and encephalopathy and long-term AED use. A similar conclusion was reached by investigators at the Sorbonne, Paris, France, who reviewed the literature and reports of ~600 cases in the last 30 years (Bulteau C et al. Epilepsy surgery for hemispheric syndromes in infants: hemimegalencephaly and hemispheric cortical dysplasia. **Brain Dev** 2013 Sep;35(8):742-7). Hemispheric surgical procedures are considered safe and can be performed from the first month of life. Residual insular cortex and contralateral MRI abnormalities are associated with persistent postoperative seizures and lack of cognitive improvement.

NEURO CUTANEOUS DISORDERS

mTOR INHIBITION AND TUBEROUS SCLEROSIS PREVENTION

Investigators at Children's Memorial Health Institute, Warsaw, Poland, report monozygotic twin sisters with tuberous sclerosis complex (TSC), one treated with the mTOR inhibitor everolimus since age 4 years. After 24-month follow-up, everolimus treatment was associated with a significant decrease in brain tumor volume, and the treated twin presents no facial angiofibroma, and no renal angiomyolipomas (AMLs).

The brain tumor in the nontreated sister is stable in size, but she has developed facial angiofibroma and renal AMLs. Early mTOR inhibition in TSC patients may prevent the development of TSC lesions and alter the natural history of the disease. (Kotulska K, Bortkowska J, Jozwiak S. Possible prevention of tuberous sclerosis complex lesions. **Pediatrics** 2013 Jul;132(1):e239-42). (Response: Dr Kotulska. k.kolulska@ezd.pl).

COMMENT. The approach to the management of tuberous sclerosis complex (TSC) has become more demanding because novel TSC-specific treatments are now available, and indications for intervention vary with age and organ system. Periodic MR imaging is advised every 1-3 years to exclude development of subependymal giant cell astrocytoma and need for treatment with everolimus, negating the need for invasive surgery. Routine EEG in asymptomatic TSC infants for the first year or two of life is increasingly accepted, with treatment initiation of vigabatrin if indicated at the earliest sign of developing hypsarrhythmia. (Krueger DA. **Curr Treat Options Neurol** 2013 July 13).

ATTENTION DEFICIT HYPERACTIVITY DISORDER AND EEG

METHYLPHENIDATE-INDUCED SEIZURE, ADHD AND ABNORMAL EEG

Investigators from Harvard Medical School, Boston, present the case of a 6-year-old girl with ADHD taking valproic acid for behavioral problems who had a generalized tonic-clonic convulsion shortly after a first dose of methylphenidate. She was known to have staring spells and an abnormal EEG with left centroparietal spikes but no history of electrographic seizures. A repeat EEG demonstrated continuous spike and slow wave during sleep. CSWS in sleep may be a risk factor for methylphenidate-induced seizure. (Sheen VL, Shankar M, Marin-Valencia I, Bridgemohan CH, Torres AR. Methylphenidate and continuous spike and wave during sleep in a child with attention deficit hyperactivity disorder. **Pediatr Neurol** 2013 Jul;49(1):54-7). (Response: Dr Torres. E-mail: Alcy.torres@childrens.harvard.edu).

COMMENT. This article corroborates previous reports of the utility of the EEG in children with ADHD and the increased risk of methylphenidate-induced seizures in patients with EEG centrotemporal (rolandic) spikes (Hemmer SA, et al. **Pediatr Neurol** 2001 Feb;24(2):99-102) and CSWS in sleep. Sleep deprivation in preparation for a sleep EEG is essential to exclude epileptiform abnormalities; one quarter of nonepileptic children with ADHD have epileptiform discharges in the sleep-deprived EEG compared to only 7% in awake only EEG, and slightly more than half discharges are focal (Millichap JJ et al. **J Child Neurol** 2011 Jan;26(1):6-11).