COMMENT. In 2003, 20 infants were hospitalized in Israel with severe neurological symptoms, including ophthalmoplegia, vomiting, nystagmus, seizures and coma. Brain MRI showed hyperintense signal in the basal ganglia, mamillary bodies and periaqueductal grey matter. Two died of cardiomyopathy and 10 had residual cardiac and brain damage. Wernicke’s encephalopathy was suspected, and examination of the infant formula found an absence of vitamin B1. Treatment with supplementary thiamine resulted in improvements in affected infants. The above study was conducted on patients fed the same thiamine-deficient formula but showing no neurological deficits. The research demonstrates the influence of diet deficiencies, specifically thiamine, on the developing brain of infants and the effect on language performance in later childhood.

SEIZURE DISORDERS

INTERHEMISPHERIC FUNCTIONAL CONNECTIVITY IN CHILDHOOD ABSENCE EPILEPSY

Using a blood oxygen level-dependent resting functional connectivity approach to analyze EEG-fMRI data, the properties of bihemispheric brain networks in 16 patients with childhood absence epilepsy (CAE) were investigated during the interictal period, in a study at Yale University School of Medicine, New Haven, CT. Resting functional connectivity between hemispheres was significantly increased in the lateral orbitofrontal cortex of patients with CAE compared to normal matched controls. The abnormal increased connectivity between hemispheres is demonstrated in patients with CAE both during and between seizures. Resting functional connectivity analysis is a promising biomarker for altered brain function in CAE during the interictal period. The findings provide further evidence for focal bilateral network abnormalities in CAE. (Bai X, Guo J, Killory B, et al. Resting functional connectivity between the hemispheres in childhood absence epilepsy. Neurology June 7, 2011;76(23):1960-1967). (Response and reprints: Dr Hal Blumenfeld, Dept Neurology, 333 Cedar St, New Haven, CT 06520. E-mail: hal.blumenfeld@yale.edu).

COMMENT. In an editorial, Zempel JM and Ciliberto M (Neurology 2011;76:1952-1953) discuss fluctuating concepts of CAE referring to the recent interest in attention, behavioral, cognitive, and linguistic impairments complicating CAE. The comparative trial of 3 AEDs for absence epilepsy (ethosuximide, valproic acid, and lamotrigine) were all effective for seizure control; ethosuximide had an additional beneficial effect, improved attention. Increased lateral orbitofrontal connectivity demonstrated in the Yale study may be a biomarker of this attention deficit-in CAE. Functional connectivity diagrams of brain networks and interictal spike/wave paroxysms may, in the future, predict the degree of cognitive impairment and lack of awareness and attention in a child with CAE.

Motor system hyperconnectivity in juvenile myoclonic epilepsy. Vollmar C et al report increased functional connectivity between the motor system and frontoparietal cognitive networks, providing an explanation for the triggering of myoclonic jerks by cognitive effort in JME patients. (Brain June 2011;134:1710-1719).