

HYPARRHYTHMIA VARIANT PATTERNS

The frequency of hyparrhythmia variant or atypical patterns was determined by analysis of pre-ACTH EEG records of 53 consecutive patients with infantile spasms at Children's Hospital, Harvard Medical School, Boston, MA. Hyparrhythmia was scored (16 most severe, 0 normal EEG) based on degree of disorganization, diffuse delta, voltage, spikes, burst suppression (BS), etc. Variant patterns occurred in 69% of records. Hemihyparrhythmia or BS was characteristic of spasms caused by cerebral dysgenesis. Sleep patterns were persistently abnormal in infants with perinatal hypoxic-ischemic encephalopathy. Lower hyparrhythmia scores (<10), <75% delta activity, and absence of electrodecremental discharges correlated with more favorable outcome, whereas variant patterns had no prognostic significance. (Kramer U, Sue WC, Mikati MA. Hyparrhythmia: Frequency of variant patterns and correlation with etiology and outcome. *Neurology* Jan 1997;48:197-203). (Reprints: Dr MA Mikati, Department of Pediatrics, c/o American University of Beirut New York Office, 850 Third Ave, New York, NY 10022).

COMMENT. Analysis of pre-ACTH EEGs in patients with West syndrome may reflect the pathophysiology and prognosis. Higher severity scores of hyparrhythmia correlate with poor outcome. Variant patterns of hyparrhythmia are frequent and show no correlation with prognosis; they should be included within the definition and indications for treatment with ACTH.

Cortical hypometabolism and delayed myelination were reported in a study of 18 children with West syndrome at Nagoya University School of Medicine, Japan. (Natsume J, Watanabe K, Maeda N et al. *Epilepsia* Dec 1996;37:1180-1184). Hypometabolism on PET scans at the onset of spasms or at 10 months is correlated with MRI evidence of delayed myelination at age 10 months.

POSITIVE SPIKES WITH NEURONAL MIGRATION DISORDERS

Five of fifteen children with histologically proven neuronal migration disorders (NMD) and refractory localization-related epilepsy had surface positive spikes or sharp waves or both on scalp EEG performed preoperatively in a study at The Hospital for Sick Children, Toronto, Canada. Patients with positive epileptiform discharges had earlier onset of seizures with focal motor pattern, poor response to AEDs, a higher frequency of hemiparesis preoperatively, and surgery at a younger age with poor outcome. Lesions involving the rolandic fissure are more extended than those seen on the MRI and cannot be completely resected. (Otsubo H, Steinlin M, Hwang PA, et al. Positive epileptiform discharges in children with neuronal migration disorders. *Pediatr Neurol* Jan 1997;16:23-31). (Respond: Dr Otsubo, EEG Laboratory, Division of Neurology, Hospital for Sick Children, 555 University Ave, Toronto, Ontario M5G 1X8, Canada).

COMMENT. Compared to the usual negative polarity of most epileptiform abnormalities, focal positive spike waves encountered with neuronal migration disorders are predictive of more extensive lesions, higher incidence of hemiparesis, focal motor seizures refractory to AEDs, and less favorable surgical outcome associated with incomplete resection.

The pathomechanism of the neuronal migration disorder seen in Miller-Dieker syndrome (smooth cerebral surface and four