

second dose has proven safe after 4 hours.

### **ACTH vs VIGABATRIN IN INFANTILE SPASMS**

The comparative efficacy and safety of ACTH (110 IU/m<sup>2</sup> once daily for 15 days) and vigabatrin (100-150 mg/kg/daily in bid doses) was evaluated by a retrospective analysis of medical records of 42 infants (21 in each group) with infantile spasms and hypsarrhythmia treated at the Université de Montreal, Hôpital Sainte-Justine, Montreal, Canada. Seizure response was similar in the ACTH and vigabatrin-treated infants at 12 month follow-up (61% vs 71% benefited, respectively), but vigabatrin was better tolerated than ACTH. Side effects of ACTH in 5 patients included hypertension and Cushing's syndrome, requiring discontinuation of therapy in 3; hypotonia and sleep disturbance in 3 vigabatrin-treated patients were transient and did not require drug withdrawal. No cases of visual field restriction were encountered. Vigabatrin was recommended as the first-line treatment for infantile spasms. (Cossette P, Riviello JJ, Carmant L. ACTH versus vigabatrin therapy in infantile spasms: a retrospective study. Neurology May 1999;52:1691-1694). (Reprints: Dr Lionel Carmant, Service de Neurologie, Hôpital Sainte-Justine, 3175, Cote Sainte-Catherine, Montreal, Qc, H3T 1C5 Canada).

COMMENT. Vigabatrin and ACTH appear to show equal effectiveness in the treatment of infantile spasms, but the plethora of reports of vigabatrin-induced visual field defects in adults are a concern. The majority of the cases cited are in adults, but asymptomatic visual field constriction also occurs in children. Two affected children, ages 10 and 15, treated with vigabatrin in doses ranging from 1000 to 3500 mg/kg, are reported from the Children's Hospital, University Hospital of Kuopio, Finland. (Vanhatalo S, Paakkonen L. Neurology May 1999;52:1713-1714).

At least with long-term therapy, vigabatrin should be used with caution in infants, until more is known about the risk factors involved.

### **PERIVENTRICULAR HETEROTOPIA AND ABSENCE EPILEPSY**

A 7-year-old child with absence seizures and a 3-Hz generalized and occasional focal, spike-and-wave EEG pattern, responsive to treatment with valproic acid, had an MRI with periventricular nodular heterotopias, primarily right sided. The neurologic examination was normal. (Giza CC, Kuratani JD, Cokely H, Sankar R. Periventricular nodular heterotopia and childhood absence epilepsy. Pediatr Neurol April 1999;20:315-318). (Respond: Dr Raman Sankar, Pediatric Neurology, Box 951752, UCLA School of Medicine, Los Angeles, CA 90095).

COMMENT. Focal abnormalities in an otherwise typical and generalized EEG pattern for childhood absence epilepsy prompted an MRI in this case which uncovered the unexpected cerebral migration lesion.

### **NOCTURNAL FRONTAL LOBE EPILEPSY**

The clinical and polysomnographic findings in 100 consecutive cases of nocturnal frontal lobe epilepsy (NFLE) were analysed in a study at the Neurological Institute, University of Bologna, Italy. Males predominated, in a male/female ratio of 7:3. Nocturnal paroxysmal episodes presented at any age from 1 to 64 years (mean 14+/-10 years) but mainly during infancy and adolescence; parasomnias in infancy were followed by NFLE seizures after intervals of 1 to 30 years. A family history of epilepsy or parasomnias occurred in 25% or 39%, respectively. Past histories included sleep disorders (talking, enuresis, head banging, and sleep walking) in early childhood; 7 had birth anoxia, 3 febrile convulsions, and 3 antecedent mild head trauma. CT or MRI

abnormalities were found in 14%. EEGs were epileptiform in only 56% of ictal and 50% of interictal sleep records; epileptiform discharges were recorded only by sphenoidal electrodes in 13%.

VideoEEG showed that NFLE is a spectrum of seizure phenomena (*paroxysmal arousal from sleep, nocturnal paroxysmal dystonia, and episodic nocturnal wanderings*), representing a continuum of different clinical manifestations of a heterogeneous epilepsy syndrome. Carbamazepine controlled the seizures completely in 16% and partially in 48%. (Provini F, Plazzi G, Tinuper P, Vandi S, Lugaresi E, Montagna P. Nocturnal frontal lobe epilepsy. A clinical and polygraphic overview of 100 consecutive cases. Brain June 1999;122:1017-1031). (Respond: Elio Lugaresi MD, Istituto di Clinica Neurologica, Universita di Bologna, Via Ugo Foscolo 7, 40123 Bologna, Italy).

COMMENT. The diagnosis of nocturnal frontal lobe epilepsy (NFLE) should be suspected in infants and children with paroxysmal nocturnal motor episodes during sleep, characterized by clusters of repetitious stereotyped movements that persist into adolescence and are associated with dystonic posturing or agitated behavior. Pelvic thrusting, facial grimacing and moaning are frequent repeated patterns of behavior during seizures. Diurnal episodes include generalized shivering with loss of consciousness, and complaints of tingling and daytime sleepiness. NFLE is often familial and autosomal dominant in inheritance. NFLE responds to treatment with carbamazepine or clonazepam, but seizures almost always recur when therapy is withdrawn.

Benign nocturnal parasomnias - nightmares, night terrors and somnambulism - may be differentiated from NFLE by a different time course, with remission within 7 years, whereas NFLE persists into adolescence. Parasomnias occur infrequently and at irregular intervals, whereas NFLE occurs nightly and repeatedly. Furthermore, parasomnias are not manifested by dystonic posturing or violent agitated motor behavior. The EEG and especially video EEG is diagnostic in approximately 50% of patients. A trial of antiepileptic drugs may sometimes be warranted on clinical grounds, in the absence of EEG confirmation. (See Progress in Pediatric Neurology III, PNB Publ, 1997;pp87-88).

## TOXIC-INFECTIOUS DISORDERS

### SEQUELAE OF HEMOLYTIC UREMIC SYNDROME

The neuropsychological function of 91 children who had recovered from hemolytic uremic syndrome (HUS), without severe neurological dysfunction (stroke, hemiplegia, blindness, retardation), and case controls admitted for non-HUS illness, was determined in a multicenter study at six tertiary care hospitals reporting to the Canadian Pediatric Kidney Disease Research Center, Ottawa, Canada. Scores on verbal ability tests were lower in patients with the highest serum creatinine concentrations during the HUS illness, but the severity of the acute renal failure was not correlated with neuropsychological measures. There was no increased risk of attention deficit disorder among patients recovering from HUS. (Schlieper A, Orrbine E, Wells GA et al. Neuropsychological sequelae of hemolytic uraemic syndrome. Arch Dis Child March 1999;80:214-220). (Respond: Dr PC Rowe, Brady 212, Johns Hopkins Hospital, 600 North Wolfe Street, Baltimore, MD 21287).

COMMENT. Children recovering from acute hemolytic uremic syndrome without serious neurologic sequelae are not at risk of learning, behavior, or attention problems. Neuropsychological tests are not required as a routine part of the follow-up, but only as symptoms and school performance dictate.