

years on medication. The change in lipid metabolism may be associated with induction of liver enzymes during carbamazepine treatment for epilepsy, and may also have clinical relevance to the increased incidence of atherosclerosis and coronary heart disease in patients with epilepsy. (Isojarvi JIT et al. Serum lipid levels during carbamazepine medication. A prospective study. Arch Neurol. June 1993; 50: 590-593). (Reprints: Dr Isojarvi, Department of Neurology, University of Oulo, SF-90220 Oulo, Finland).

COMMENT. The data support the hypothesis that the increase in serum cholesterol levels during carbamazepine therapy for epilepsy may be associated with liver enzyme induction. The changes in serum lipids may also be a consequence of hormonal effects, notably a decrease in free thyroxine, observed with carbamazepine. Patients receiving carbamazepine should have base line lipid profiles and serum lipid monitoring. Carbamazepine may not be an adjunct drug of choice in children receiving the ketogenic diet for refractory epilepsy.

An elevation of very long chain fatty acids found in 13 of 22 plasma samples from patients on a standard ketogenic diet for uncontrolled seizures may lead to diagnostic confusion with peroxisomal disorders. Valproic acid was taken by 5 of the patients tested. (Theda C et al. J Pediatr May 1993; 122: 724-6).

JUVENILE MYOCLONIC EPILEPSY

The clinical and EEG features and reasons for frequent misdiagnosis of juvenile myoclonic epilepsy are reviewed by the Epilepsy Research Group, Institute of Neurology, National Hospital, and St Thomas' Hospital, London, England. The triad of absence seizures, myoclonic jerks, and GTCs shows a characteristic age-related onset. Absence seizures begin between 5 and 16 years, myoclonic jerks follow about 4 years later, usually around age 15 years, and GTCs appear within a few months after the myoclonic jerks. Myoclonic jerks and GTCs occur mainly on awakening. Apart from tremor, the neurologic exam is normal. Photosensitivity is present in 50% of patients, and seizures are precipitated by sleep deprivation, fatigue, alcohol, anxiety, and hyperventilation. Treatment with valproic acid is usually effective, and may be supplemented with clonazepam at bedtime if necessary. Lifelong anticonvulsant treatment is required, and withdrawal of medication may result in status epilepticus. (Grunewald RA, Panayiotopoulos CP. Juvenile myoclonic epilepsy. A review. Arch Neurol June 1993; 50: 594-598). (Reprints: Dr Grunewald, Epilepsy Research Group, Institute of Neurology, National Hospital, Queen Square, London, England WC1N 3BG).

COMMENT. A high index of suspicion for JME is indicated in young patients with early-morning seizures, especially those poorly controlled

with carbamazepine or associated with sleep deprivation or alcohol consumption.

Diagnosis of JME was delayed by a mean of 14 years in 15 patients identified among 180 referrals to a new epilepsy clinic at St Thomas' Hospital, London, a prevalence of 8.3%. At least 11 had been examined previously by a neurologist, and 7 had received inappropriate anticonvulsants. (Grunewald RA et al. J Neur Neurosurg & Psychiat 1992; 55:497).

BRAIN TUMORS

CHEMOTHERAPY, DELAYED RADIATION AND BRAIN TUMORS

To avoid the severe irradiation-related neurotoxic effects of postoperative treatment of malignant brain tumors in infants and children less than 3 years of age, the Pediatric Oncology Group, centered at St Louis, MO, conducted a prospective study of primary postoperative chemotherapy. Treatment consisted of two 28-day cycles of cyclophosphamide plus vincristine, followed by one 28-day cycle of cisplatin plus etoposide. The sequence was repeated 1) until the tumor showed progression or 2) for 2 years in 132 children < 24 months of age at diagnosis and 3) for 1 year in 66 children of 24-36 months of age. Chemotherapy was then followed by radiation. The first two treatment cycles produced complete or partial responses in 39% of 102 patients evaluated. Medulloblastomas, malignant gliomas, and ependymomas were most responsive, whereas brain-stem gliomas or embryonal tumors showed little or no response. The progression-free survival was 40% at the completion of treatment schedules. Cognitive evaluations at base line and after 1 year of chemotherapy showed no deterioration. (Duffner PK et al. Postoperative chemotherapy and delayed radiation in children less than three years of age with malignant brain tumors. N Engl J Med June 17 1993; 328: 1725-31). (Reprints: Dr Duffner, Pediatric oncology Group Operations Office (8633, 8634), 4949 W Pine Blvd, St Louis, MO 63108).

COMMENT. Chemotherapy prevents disease progression and permits radiation therapy to be delayed for one to two years in about 40% of infants and very young children with malignant brain tumors. The best results are obtained in those with total surgical resection of localized disease. Chemotherapy is least effective in children with embryonal, primitive neuroectodermal tumors. Dr JC Allen, New York University Medical Center, cautions that delaying cerebral radiotherapy for 1 to 2 years may not spare an infant neurocognitive complications, and that intensive chemotherapy may also prove to have a detrimental effect on brain development. (Editorial. N Engl J Med June 17 1993; 328: 1780-1).