

COMMENT. The authors conclude that excisional surgery can be performed safely in selected infants with medically uncontrolled malignant partial seizures and may improve long-term seizure outcome. They emphasize referral to a center specializing in early childhood epilepsy surgery. The same authors report at the 42nd Annual Meeting of the AAN that intractable focal seizures in childhood have a histopathological spectrum distinct from that of adults. Malformations, particularly neuronal migration disorders are most frequent in infants whereas hippocampal sclerosis, a common pathology in adult epileptics, did not occur in the infants or the children in this report. (Neurology April 1990; 40 (Suppl I):187).

ANTERIOR TEMPORAL LOBECTOMY IN REFRACTORY COMPLEX PARTIAL SEIZURES

The outcome of 22 patients with onset of complex partial seizures (CPS) in early childhood and treated by anterior temporal lobectomy after intervals varying from three to 28 years, is reported from the Epilepsy Research Center, Baylor College of Medicine, Houston, TX. All patients showed improved seizure control, the majority having a greater than 95% reduction in seizure frequency. Psychosocial, behavioral, and educational problems occurred more frequently in patients whose surgery was delayed until adult life. Neuropathologic abnormalities were found in both the mesial and lateral portions of the temporal lobe. Mesial abnormalities included the classical Ammon's horn sclerosis and ganglioglioma. All the brain specimens showed congenital malformations or "microdysgenesis". The authors considered surgery, performed soon after medical intractability has been determined, may limit the problems associated with prolonged uncontrolled seizures. (Mizrahi EM et al. Anterior temporal lobectomy and medically refractory temporal lobe epilepsy of childhood. Epilepsia May/June 1990; 31:301-312).

COMMENT. In these patients with seizure onset between two and ten years of age Ammon's horn sclerosis occurred in 16 of the 22 patients. This finding contrasted with the absence of hippocampal sclerosis in patients with seizures beginning in infancy. (See Duchowny et al. Neurology 1990; 40:980).

VALPROATE, CARNITINE, AND LIPID METABOLISM

The effects of valproate (VPA) on carnitine and lipid metabolism and on liver function were assessed in 213 outpatients from five centers and reported from the Instituto di Ricerche Farmacologiche "Mario Negri," Milan, Italy. The mean total and free carnitine levels were significantly lower in patients on polytherapy. A significant correlation was found between serum ammonia levels and VPA dosage. VPA monotherapy and polytherapy were associated with significantly elevated cholesterol levels, especially "HDL". The authors concluded that impairment of carnitine metabolism and liver function by VPA does not appear to be a clinically important phenomenon especially when VPA is administered as monotherapy to well nourished patients. There was no correlation between carnitine deficiency and reports of

anticonvulsant clinical toxicity, e.g. somnolence, behavioral disturbance, headache, increased appetite, weight gain, anorexia, ataxia, and tremor. (Beghi E et al and the Collaborative Group for the Study of Epilepsy. Valproate, carnitine metabolism, and biochemical indicators of liver function. Epilepsia May/June 1990; 31:346-352).

COMMENT. These data confirm previous findings that VPA impairs carnitine metabolism. In contrast to other reports, this study showed a significant correlation between serum ammonia and VPA dosage. The observed change in lipid metabolism with increased cholesterol levels during treatment with VPA and other anticonvulsants is important in the evaluation of children receiving the ketogenic diet as a supplement to anticonvulsant drugs. Carnitine deficiency has been found in hyperlipemic patients and carnitine replacement therapy may correct the hyperlipemia. Children, especially females and younger males, should be tested for carnitine deficiency during treatment with anticonvulsant drugs and particularly with VPA polytherapy.

TREATMENT OF INFANTILE SPASMS

The rationale, dosage, and side effects of ACTH treatment of infantile spasms are reviewed from the Department of Neurology, University of Southern California School of Medicine, and Children's Hospital of Los Angeles, CA. Admission to hospital is recommended for the initiation of ACTH therapy so that baseline laboratory tests can be obtained and response to treatment can be supervised for the first three days. The author recommends an initial dose of ACTH 150 units/m²/day (Acthar gel), 80 units/ml intramuscularly in two divided doses for one week. In the second week the dose recommended is 75 units/m²/day in one daily dose for one week. In the third week the dose is 75 units/m² every other day for one week. ACTH is gradually withdrawn over the next nine weeks. A change in the lot number of ACTH gel is indicated when no response occurs in two weeks. The author admits controversy concerning the dose, duration of therapy, and effectiveness of ACTH relative to oral steroids as well as the long-term benefit of either of these therapies. (Snead OC III. Treatment of infantile spasms. Pediatr Neurol May/June 1990; 6: 147-150).

COMMENT. Exceptionally large doses of ACTH recommended in this report were also used by Riikonen R and Simell O in the treatment of infantile spasms associated with tuberous sclerosis. (See Ped Neur Briefs April 1990; 4:30-31). There was an unusually high incidence of side effects, including arterial hypertension, with two patients developing cardiac failure and three with fluid retention in polycystic kidneys. My own preference is for smaller doses (10-20 units of Acthar gel daily by intramuscular injection for three weeks) with the avoidance of cushingoid complications and a lesser incidence of other side effects. Early treatment is important in terms of the response to therapy and possibly in relation to subsequent development.