

epilepsy. Neurology April 1994;44 (Suppl 2):A273 (abstr). (Respond: Dr Edwin Trevathan, 5455 Meridian Mark Rd, Ste 530, Atlanta, GA).

COMMENT. Clinically significant weight loss and anorexia were troublesome side effects during a trial of felbamate in 68 children and adults with intractable seizures at Rush-Presbyterian-St Luke's Medical Center, Chicago. (Waicosky K et al. Weight loss in patients taking felbamate. Neurology April 1994;44 (Suppl 2):A296). Insomnia was reported in 25% of felbamate-treated patients in a further study of 16 patients (Luciano D et al. Neurology April 1994;44 (Suppl 2):A296).

Gabapentin (Neurontin), another newer anticonvulsant, recently approved and introduced for treatment of partial and secondarily generalized seizures in children older than 12 years and in adults, was found to be safe and well-tolerated as monotherapy in a multicenter study. (Hayes A et al. An open-label multicenter study of gabapentin (Neurontin) monotherapy and safety in medically refractory patients with partial seizures. Neurology April 1994;44 (Suppl 2):A204). Adverse effects have been minor and psychometric testing revealed no cognitive impairments. In follow-up studies > 1 year, there was no evidence of chronic toxicity. Unlike other anticonvulsants, gabapentin is not metabolized by the liver and is free of interactions with other drugs. (Leppik IE. Epilepsia 1994;35 (Suppl 4):S29-S40).

PSEUDOTUMOR CEREBRI

SYMPTOMS AND SIGNS OF PSEUDOTUMOR

A review of charts of 30 children with idiopathic intracranial hypertension, seen in a 30 year period between 1960 and 1990, is reported from the University of Iowa Hospital and Clinics. Common presenting symptoms included headache (63%), vomiting (43%), diplopia (36%), blurred vision (26%), and nausea (23%). The majority were heavier than the 50th percentile. All had bilateral papilledema, 30% had retinal hemorrhage, and 53% enlarged blind spots. CTs and MRIs were normal. Associated illnesses were URIs in 16%, otitis in 16%, head trauma in 6%; 46% were otherwise healthy. Six had taken antibiotics, and 4 were on vitamins, including vitamin A. Neurologic signs included VIth nerve palsy in 14, and ataxia in 4. Steroids were used in 9 and acetazolamide in 8. (Babikian P et al. Idiopathic intracranial hypertension in children: The Iowa experience. J Child Neurol April 1994;9:144-149). (Respond: Dr James J Corbett, Dept Neurology, UMC, 2500 North State Street, Jackson, MS 39216).

COMMENT. Abnormalities on neurologic examination are more common in children than in adults. The infrequent occurrence or recognition of the syndrome in infants and young children is noteworthy. A review of the literature before 1960 and prior to the period examined in the Iowa study showed that 84 (37%) of 224 patients were children and of these 75 (90%) were between 5 and 15 years of age. (Millichap JG. Benign intracranial hypertension and otitic hydrocephalus. Pediatrics Feb 1959 ;23:257). Antecedent otitis media was reported in 65 (29%), and mild head injury or infection other than otitis in 66 (29%). The 93 (41%) cases classified as idiopathic occurred principally in adults in this earlier series whereas approximately one half the pediatric cases in the later

Iowa report were unassociated with otitis or other infection, trauma, or predisposing illness. The advent of antibiotics and virtual abolition of mastoiditis and "otitic hydrocephalus" accounts for the change in frequency of predisposing causes and the increase in "idiopathic" cases. For a recent major review of pediatric pseudotumor cerebri, see Lessell S. Survey of Ophthalmology 1992;37:155-166).

Combined therapy with acetazolamide (37 - 100 mg/kg/daily) and furosemide (1 mg/kg/daily) was effective in treating raised intracranial pressure in 8 children with pseudotumor cerebri at the Department of Pediatrics, University of Stellenbosch and Tygerberg Hospital, Republic of South Africa (Schoeman JF. J Child Neurol April 1994;9:130-134). Repeated lumbar cerebrospinal fluid pressure monitoring was used to evaluate response to therapy, but clinical monitoring correlated well and would be adequate in most children.

RETT SYNDROME (RS)

ACYL-COA DEHYDROGENASE DEFICIENCY AND RS

A female infant with medium-chain acyl-CoA dehydrogenase (MCAD) deficiency who was diagnosed with Rett syndrome at 3.5 years is reported from Twenteborg Hospital, Almelo, and Wilhelmina Kinderziekenhuis, Utrecht, The Netherlands. At 13 months her development was normal. By 20 months she could not walk, her language development had ceased, and tremor with loss of purposeful hand movements was noted. At 30 months she had hypotonia, increased tremor and "handwashing" movements. At 3 years she was mentally retarded with autistic features, and the EEG showed bilateral spikes and spike wave activity and a slow waking background rhythm. The head circumference was at the 98th percentile from birth to 17 months and 50th percentile at 52 months. Four additional Rett syndrome patients had normal lymphocyte MCAD assays. (Beekman RP et al. Rett syndrome in a patient with medium chain acyl-CoA dehydrogenase deficiency. Eur J Pediatr April 1994;153:264-266). (Respond: Dr RP Beekman, Wilhelmina Kinderziekenhuis, PO Box 18009, 3501 CA Utrecht, The Netherlands).

COMMENT. The authors found no reason to propose a causal relationship between MCAD deficiency and Rett syndrome.

A controlled study of an oral opiate antagonist, Naltrexone, in 25 patients with Rett syndrome at the University of Alabama, Birmingham, AL, and other centers, showed a beneficial effect on respiratory irregularities and improved oxygenation but negative effects on development measured by Bayley scales. (Percy AK et al. Ann Neurol April 1994;35:464-470). The hypothesis that naltrexone may be beneficial in Rett syndrome followed from reports of elevated levels of B-endorphins in the CSF of Rett syndrome patients. Further, the intraventricular administration of endorphins in animals produces naloxone-reversible signs similar to those of Rett syndrome.