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 dehyrogenase (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 intraventicular administration of endorphins in animals produces naloxone-reversible signs similar to those of Rett syndrome.