SEIZURE DISORDERS

PROGNOSIS OF INFANTILE SPASMS (WEST SYNDROME)

Investigators at the Instituto di Neuropsichiatria Infantile, University of Rome, Italy, followed 58 cases of West syndrome for 3 years or more (mean 5 yr 5 mo). Eight (14%) were classified as idiopathic, and 50 (86%) were symptomatic, of which 17 were secondary to birth asphyxia or cerebral hemorrhage and 11 (19% of total) had tuberous sclerosis. All were treated with ACTH, 25-50 IU daily for 3 weeks and 25 IU daily for further 3 weeks or more if no response.

IQ was normal or borderline (>70) in 5 (8%) and retarded in 53 (92%). A low IQ at final follow-up was correlated with developmental and/or neurological abnormalities before the onset of spasms, symptomatic etiology, and abnormal CT findings (cerebral atrophy 30 (52%), calcifications 11 (19%), callosal agenesis 3 (5%). An IQ above 50 in 13 (23%) was associated with early speech development (first word before 2 years and two-word sentence before 3 years) and an IQ below 50 was found in children with retarded speech development. Motor milestones did not correlate with subsequent mental development even in cases without neuromotor impairment. Whereas clinical and CT evidence of cerebral damage correlated with mental retardation, the absence of cerebral abnormalities was not predictive of a normal mental development. A poor prognosis in WS was predictable but normal development at follow-up was not reliably associated with favorable variables at onset. (Favata I, Leuzzi Y, Curatolo P. Mental outcome in West syndrome: prognostic value of some clinical factors. J Ment Def Res 1987; 31: 9-15).

COMMENT: Of 1,558 cases of West syndrome reported in the literature between 1954 and 1973, a 20-year span, 624 (40%) were idiopathic and 934 (60%) were symptomatic (Lacy JR, Penry JL. Infantile Spasms. Raven Press, New York, 1976). The unusually high percentage of symptomatic cases in this study could explain the relatively higher incidence of mental retardation. Could the 52% incidence of cerebral atrophy demonstrated by CT be caused in part by ACTH? As the authors point out, their data related to a poor mental outcome cannot strictly be regarded as prognostic factors since they are an expression of the symptomatic character of WS, accounting for 86% of their cases. Among 11 patients with tuberous sclerosis, 4 (36%) had an IQ>50 at follow-up and 7 (64%) were moderately to severely retarded. Unfortunately, the relation of IQ to the age at time of treatment and response to ACTH was not evaluated, a frequently debated question. In a study at the Mayo Clinic (Millichap JG, Bickford RG. JAMA 1962; 182:523), psychomotor retardation was generally more severe in patients with symptomatic infantile spasms than in those with cryptogenic seizures. ACTH was possibly more effective in patients with IQ's of 70 or above than in those with retarded development before treatment. Control of infantile spasms and hypsarrhythmia by ACTH was approximately 50% and equal in cryptogenic and symptomatic groups of patients.

TOXICITY OF NITRAZEPAM (MOGADON)

A pediatric neurologist and his associates at the Children's Mercy Hospital, Kansas City, report 6 deaths among 80 patients with intractable epilepsy treated with nitrazepam. The patients who died were 13-39 mos of age (mean 28 mos) and had received nitrazepam for 2 to 19 months (mean 8 mos) in a dosage of 0.9-2.7 mg/kg/d (mean 1.4). Up to 2 additional but unnamed antiepileptic