

HIGH-DOSE STEROIDS IN RASMUSSEN'S SYNDROME

Ten of 17 patients with Rasmussen's syndrome receiving IV methylprednisolone and/or oral prednisolone, and eight of nine patients receiving immunoglobulins showed some short-term reduction in seizure frequency in a multicenter international report. Side effects included fluid retention, psychotic symptoms and behavior problems. The authors propose a central register and standardized protocol with initial trial of IV immunoglobulin (400 mg/kg/d on 3 successive days) followed by monthly one day treatments if improvement occurs. Steroid therapy (IV methylprednisolone, 400 mg/m² on 3 alternate days followed by monthly single infusions for one year or longer, and oral prednisolone starting at 2 mg/kg/d) is recommended for patients not responding to IVIG. (Hart YM, Cortez M, Andermann F et al. Medical treatment of Rasmussen's syndrome (chronic encephalitis and epilepsy): Effect of high-dose steroids or immunoglobulins in 19 patients. Neurology June 1994;44:1030-1036). (Respond: Dr F Andermann, Montreal Neurological Institute, 3801 University Street, Montreal, PQ, Canada H3A 2B4).

COMMENT. The patients were treated at the Montreal Neurological and Children's Hospitals; the National Hospital, London; Great Ormond Street Hospital, London; Hospital for Sick Children, Toronto; and Hospital Juan Garrahan, Buenos Aires. The diagnosis was confirmed by biopsy in all but three of the patients. As with infantile spasms, the earlier the therapy the better the results. Since ACTH is usually considered superior to prednisone in the treatment of infantile spasms, it is surprising that only one of the 17 patients with Rasmussen's syndrome received ACTH and ACTH is omitted from the recommended treatment protocol for further trials. If a comparison with infantile spasms is carried further, patients benefited by steroids or ACTH usually respond within 4 to 8 weeks and prolongation of therapy in non-responders is generally ineffective and is accompanied by serious toxicity. The authors recommend frequent monitoring of patients on high-dose steroids when continued for the suggested periods of 1 to 2 years or longer and caution that improvements may be delayed for several months.

CARBAMAZEPINE-RELATED STATUS EPILEPTICUS

High serum carbamazepine-epoxide concentrations were correlated with unexpected seizure exacerbation and partial status epilepticus in 6 young adults reported from the Marshfield Clinic, WI, and the Mayo Clinic, MN. All patients were mentally retarded. Ages at epilepsy onset ranged from 1 month to 11 years. Seizure exacerbation coincided with changes in drug combinations other than CBZ: VPA dosage had recently been increased in 4 patients and phenytoin had been discontinued in 1 who also took felbamate. CBZ dosage and serum levels had been therapeutic and stable for 1 to 14 years, whereas CBZ-10,11-epoxide levels exceeded an upper limit of 4 mcg/ml. CBZ-epoxide/CBZ ratios were greater than the accepted 0.2 in patients on polytherapy. Withholding CBZ was followed by seizure control within 2 to 3 days, and CBZ-epoxide toxicity (lethargy and ataxia) resolved. Withdrawal of VPA was also corrective in 1 patient who continued CBZ. (So EL et al. Seizure exacerbation and status epilepticus related to carbamazepine-10,11-epoxide. Ann Neurol June 1994;35:743-746). (Respond: Dr So, Epilepsy Service, Neurology, Mayo Clinic, 200 1st Street SW, Rochester, MN 55905).