HIGH-DOSE STEROIDS IN RASMUSSEN'S SYNDROME

Ten of 17 patients with Rasmussen's syndrome receiving IV methylprednisolone and/or oral predisolone, 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/m^2 on 3 alternate days followed by monthly single infusions for one year or longer, and oral predisolone 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).

COMMENT. CBZ-epoxide serum levels should be measured in carbamazepine-treated patients with unexplained seizure exacerbation or toxicity. Risk factors for CBZ-epoxide induced status and toxicity include high dose CBZ, combination therapies, and patients with mental retardation who often require polytherapy. Valproate, primidone, and felbamate combined with CBZ may increase levels of CBZ-epoxide by altering metabolic conversion or breakdown. In contrast, phenytoin promotes the conversion of the epoxide into an inactive form and reduces risk of CBZ-related toxicity and seizure exacerbation. Gabapentin, an AED having no drug interactions, should reduce the risk of these complications when polytherapy is considered essential.

PROPOFOL ANESTHESIA-INDUCED SEIZURES

A case of a healthy young man who developed seizures and generalized paroxysmal fast activity in the EEG following use of propofol for anesthesia in minor surgery is reported from the Department of Neurology, University of South Alabama, Mobile, AL. Myoclonic jerking and obtundation developed shortly after anesthesia and a generalized seizure associated with EEG paroxysmal fast activity and controlled with divalproex occurred the following day. (Nowack WJ, Jordan R. Propofol, seizures and generalized paroxysmal fast activity in the EEG. Clin Electroencephalogr July 1994;25:110-114). (Reprints: William J Nowack MD, Dept of Neurology, Univ of South Alabama, Mobile, AL 36617).

COMMENT. Propofol anesthesia has been associated with seizures, myoclonic jerking and opisthotonic posturing. Spikes, polyspikes and spike wave complexes in the EEG have also been reported. The fast activity in the EEG in the above case was considered to be epileptiform, resolving after short-term anticonvulsant therapy.

SEIZURE DISORDERS

CORTICAL HYPOMETABOLISM IN WEST'S SYNDROME

The association between changes on serial PET studies and the clinical course of 12 patients with newly diagnosed West's syndrome is reported from the Departments of Pediatrics and Radiology, Nagoya University School of Medicine, Nagoya, Japan. PET with FDG revealed diffuse or focal cortical hypometabolism in 11 patients, whereas MRI showed abnormalities in only 5. In 7 patients with normal findings on a second PET, spasms ceased after treatment, whereas in 5 with persistent abnormalities on PET, spasms persisted or recurred or partial seizures developed. Patients with normal MRI and normal second PET studies had normal psychomotor development. (Maeda N, Watanabe K, Negoro T et al. Evolutional changes of cortical hypometabolism in West's syndrome. Lancet June 25, 1994;343:1620-23). (Dr Norihide Maeda, Dept of Pediatrics, Nagoya Univ Sch of Medicine, 65 Turuma-cho, Showa-ku, Nagoya 466, Japan).

COMMENT. PET has been found of value in the preoperative evaluation of patients with West's syndrome (Chugani HT et al.Ann Neurol 1990;27:406-13). It may also be used in the assessment of prognosis. See also Ped Neur Briefs March 1992 and Progress in Pediatric Neurology II, Chicago, PNB Publ, 1994, for further reference to PET and infantile spasms.