COMMENT. Fetal valproate syndrome is related to the valproate dosage in the first trimester, while withdrawal symptoms are related to the dose and free fraction plasma concentration in the third trimester. These serious complications of valproate therapy for epilepsy during pregnancy occur during monotherapy as well as polytherapy. If the use of valproate during pregnancy cannot be avoided, despite the relatively high risk of teratogenicity, and especially spina bifida, the dosage and blood levels must be meticulously monitored and maintained as low as possible. (Omtzigt JGC, Lindhout D et al. Neurology 1992;42(suppl 5):119-125).

NEUROMUSCULAR DISEASES

JUVENILE MYASTHENIA GRAVIS: DIAGNOSIS AND OUTCOME

The usefulness of various diagnostic tests, treatment, and outcome in 27 juvenile myasthenics seen over a 25 year period are reported from the Departments of Pediatrics, Neurology, and Anatomy, University of Iowa. The age of onset in 56% was after 10 years of age. One fourth had ocular myasthenia, one fourth presented with ocular signs and progressed to generalized myasthenia, and half had generalized myasthenia from onset. Ptosis, the most common presenting sign (81%), was unilateral in 33%. In order of descending frequency, the other presenting signs were generalized weakness, dysphagia, diplopia, facial weakness, dysarthria, ophthalmoplegia, and nasal speech. One patient's mother had MG. The comparative yield of tests showed positive neostigmine and edrophonium tests in 92%, serology was positive for acetylcholine receptor-binding antibodies in 63%, and repetitive distal nerve stimulation showed decrement in 33%. The yield of serology and nerve stimulation tests increased with generalization of myasthenia and when proximal nerves were also tested. Ocular myasthenics responded to pyridostigmine bromide monotherapy, while generalized myasthenia required additional medical and/or surgical therapy. Patients receiving corticosteroids had thymectomy at a later date. Those with normal thymus had a greater chance of remission without medication than patients with thymic hyperplasia. (Afifi AK, Bell WE. Tests for juvenile myasthenia gravis: comparative diagnostic yield and prediction of outcome. J Child Neurol Oct 1993;8:403-411). (Respond: Dr Afifi, Division of Child Neurology, Department of Pediatrics, University of Iowa Hospitals and Clinics, Iowa City, IA 52242).

COMMENT. Thirty-eight percent of these patients were in complete remission and without drugs when followed for more than 4 years (range, days to 20 years). The outcome was better for ocular than generalized myasthenia, 57% cf 30% in remission without drugs. A lower rate of remission in thymectomized (35%) compared to non-

thymectomized (43%) patients was a reflection of disease severity and poor response to medication.

In a report of 35 juvenile myasthenics treated at the Massachusetts General Hospital, 86% of 21 who underwent thymectomy had complete or partial remission compared to 93% of 14 treated only with medication. A higher rate of complete remission and drug withdrawal was obtained in the thymectomized group, however (29% v 14%). (Millichap JG, Dodge PR. Neurology 1960;10:1007). The earlier the surgery, the better the chance of remission (see Ped Neur Briefs April 1992;6:30).

SEIZURE DISORDERS

TEMPORAL LOBE EPILEPSY: ELECTROCLINICAL FEATURES

Results of video-EEG, PET, MRI, and pathological findings in 14 children aged 16 months to 12 years who were seizure-free after temporal lobectomy are reported in a multicenter, international study at Cleveland, Singapore, Naples, and Helsinki. In 4 children with mesiotemporal sclerosis and in 1 with cortical dysplasia, EEG showed anterior/inferior temporal interictal sharp waves and unilateral temporal seizure onset. In 9 with low- grade temporal neoplasms, the EEG findings were complex, including multifocal interictal sharp waves or falsely lateralized EEG seizure onset. Video-EEG recording was most useful for localization of the epileptogenic zone for resection in those without tumors. In patients with tumors, video-EEG confirmed the epileptic nature of the complex partial seizures but was of less localizing value. (Wyllie E et al. Temporal lobe epilepsy in early childhood. Epilepsia Sept/Oct 1993;34:859-868). (Reprints: Dr E Wyllie, Pediatric Epilepsy Program, Cleveland Clinic Foundation, 9500 Euclid Ave, Cleveland, OH 44195).

COMMENT. Three of 4 patients in this study with mesiotemporal sclerosis had a history of infantile compex febrile seizures compared with none of 10 with tumor or cortical dysplasia. An excellent outcome after temporal surgical resection in a group of 19 patients with mesiotemporal sclerosis following prolonged febrile convulsions is reported from the Montreal Neurological Institute (Abou-Khalil B, Andermann F et al. Epilepsia Sept/Oct 1993;34:878). The response and seizure control in this group was significantly better than in those without antecedent febrile convulsions.

In 15 children with temporal lobe epilepsy, aged 7-14 years, studied at the Royal Children's Hospital, Melbourne, Australia, ictal SPECT provided reliable lateralizing information to confirm that obtained from surface EEG and MRI. In 4, ictal SPECT was superior to