Canada).

COMMENT. Idiopathic intracranial hypertension is not "benign" and is associated with significant morbidity and short- or long-term symptoms, often resistant to therapy. None of the proposed therapies is of proven efficacy, and this extensive case series and review emphasizes the need for prospective studies.

NEUROMUSCULAR DISORDERS

PRESYNAPTIC CONGENITAL MYASTHENIC SYNDROME

Three patients (ages 7, 9, and 14 years) with a new form of presynaptic congenital myasthenic syndrome (CMS) are reported from the University of California, Davis; University of Minnesota; and the University of Chicago. This CMS was characterized by decreased quantal release with normal amplitude miniature end-plate potentials (MEPP), normal size of nerve terminals, and normal number of synaptic vesicles. Symptoms had presented with scoliosis at age 5 years in patient 1, delay in walking and easy fatigability at 17 months in patient 2, and as an infant with hypotonia and motor developmental delay in patient 3. Clinical findings included muscle weakness and fatigability, respiratory crisis, nystagmus (1 case), bulbar deficit, scoliosis - severe in one patient, and mild ataxia. No patient had ophthalmoplegia or mental delay. A similar disorder was reported in a close relative of patient 2, but patients 1 and 3 had no family history of neurologic disease. Electrodiagnostic evidence of abnormal neuromuscular transmission was obtained in all patients. Intracellular microelectrode studies showed a dramatic reduction of the endplate potentials (EPP) quantal content, indicative of presynaptic failure. Screening of reported pathogenic mutations in the CACNA1A and a mutational analysis of AChR subunit genes were negative. Treatment with prednisone and pyridostigmine was ineffective, while a combination of pyridostigmine and 3,4-diaminopyridine reduced the frequency of respiratory crises and resulted in improved muscle strength and exercise endurance in one patient. The deficiency of quantal release of neurotransmitter underlying this form of presynaptic CMS may be explained by an abnormal calcium metabolism or impaired endocytosis and recycling of synaptic vesicles. (Maselli RA, Kong DZ, Bowe CM et al. Presynaptic congenital myasthenic syndrome due to quantal release deficiency. Neurology July (2 of 2) 2001;57:279-289). (Reprints: Dr Ricardo A Maselli, UC Davis, 1515 Newton Ct, Rm 510, Davis, CA 95616).

COMMENT. Presynaptic congenital myasthenic syndrome (CMS) results from a deficiency in release of neurotransmitter from the nerve terminal. Familial infantile myasthenia (FIM) and a CMS associated with paucity of synaptic vesicles (PSV) have been fully described, and some additional isolated cases of presumed CMS have been reported. The molecular genetic defect for CMS has not been elucidated. In the 3 cases reported here due to quantal release deficiency, involvement of CACNA1A mutations is considered most likely and deserves further evaluation.

For further review of various types of congenital myasthenic syndromes, see Engel AG et al. 1993; and Progress in Pediatric Neurology III, 1997;pp346-7.

RECOVERY FOLLOWING NEONATAL BRACHIAL PLEXUS PALSY

The value of detailed strength testing monthly, up to 6 months of age, in predicting complete recovery was determined in a prospective study of 80 infants with brachial plexus injury followed at the Brachial Plexus Palsy Center, St Louis

Children's Hospital, MO. Strength was determined using the British Medical Research Council Scales: 0 = no contraction, 1 = trace contraction, 2 = active movement with gravity eliminated, 3 = active movement against gravity, 4 = active movement against gravity and resistance, 5 = normal. Range of motion and developmental skills were also assessed monthly up to 6 months, and then every 3 to 6 months, until complete recovery or absence of further improvement to age 2 years. Therapy was conducted at home and at the center.

The serial clinical examinations clearly differentiated patient outcomes: 53 (66%) showed complete recovery, 9 (11%) had mild weakness (4/5 strength), 7 (9%) moderate weakness (3/5 strength), and 11 (14%) severe weakness (0-2/5 strength). The permanently disabled infants were followed for a mean of 4.4 years. All of the infants with complete recovery had developed antigravity strength in biceps, triceps, and deltoid muscles by 4.5 months of age, and the majority by 3 months. Distal weakness resolved more rapidly, by 4 to 8 weeks. Of the 11 with severe residual disability, none had better than 2/5 strength in proximal muscles at age 6 months, and 64% had 0-1/5 strength in wrist and fingers. (Noetzel MJ, Park TS, Robinson S, Kaufman B. Prospective study of recovery following neonatal brachial plexus injury. J Child Neurol July 2001;16:488-492). (Respond: Dr Michael J Noetzel, One Children's Place, Room 12E25, St Louis, MO 63110).

COMMENT. In infants with neonatal brachial plexus palsy, a detailed clinical examination with strength testing up to 6 months of age may distinguish the two-thirds who will make complete recovery from the 14% of infants left with a permanent severe disability. The early identification of those with a poor outcome may help in selection of patients for surgery at or after 6 months. In some studies, recovery has continued up to 9 months, and surgical intervention at an earlier age would be advised only rarely, with total plexus lesions.

The outcome in this study is at variance with that of 149 patients followed and treated conservatively at the Children's National Medical Center, Washington, DC. (Eng GD et al. 1996; see Progress in Pediatric Neurology III, 1997;357-9). Only 6 (4%) showed complete recovery, and 92 (62%) had mild impairments of strength and function.The initial clinical impairment ratings at <3 months of age correlated closely with later exams and with serial electrodiagnostic studies. Selection criteria for surgery were not clearly defined.

Strombeck C et al, at Stockholm, recommend that surgery for OBP should be delayed until after 6 to 9 months. They examined the functional outcome at age 5 years of 247 children with OBP, with or without microsurgical reconstruction. The only benefit of operation was an improved shoulder range of movement, when compared to non-operated patients. Outcome was not correlated with timing of operation, before or after 6 months. (see Ped Neur Briefs April 2000;14:25-26).

PREDICTORS OF OUTCOME IN GUILLAIN-BARRE SYNDROME

Clinical and electrophysiologic predictors of outcome in 27 children with Guillain-Barre syndrome were determined by a retrospective review of records at the University of Iowa Hospitals and Clinics, Iowa City, IA. Patients were divided into 2 groups; Group 1, recovery (complete or partial) after more than 2 months, and group 2, within 2 months of onset. Age at presentation ranged from 1 to 16 years (mean 9 years). Males outnumbered females 2:1. Antecendent infection (within 1 month of onset) occurred in 70%; cytomegalovirus infection was present in 26%. Numbness was the the most common presenting symptom, occurring in 48%. Cranial nerve involvement in 70% (mainly VII (55%), some multiple (29%)) occurred especially in group 1. Intubation was required in 36% of