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J. GORDON MILLICHAP, M.D., F.R.C.P., EDITOR

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NEUROMUSCULAR DISORDERS

CHRONIC INFLAMMATORY NEUROPATHIES

The clinical characteristics, response to therapy, and long-term prognosis in 13 children (1.5 to 16 years of age) with chronic inflammatory demyelinating polyneuropathy (CIDP) were reviewed from records of patients seen at Washington University Medical Center, St Louis, MO, and the Royal Children's Hospital, Melbourne, Australia, between 1979 and 1994. Boys were affected more often than girls in a ratio of 1.6:1. Antecedent events noted within one month of onset occurred in 7 children (54%), and included vaccinations (measles-mumps-rubella immunization in 2), intercurrent infections (URI or tonsillitis in 4), and chicken pox in 1. Lower extremity weakness, associated with difficulty in walking, was the most common presenting symptom, found in 85% of children. Motor symptoms predominated, but sensory symptoms were also noted by 85%. Deep tendon reflexes were diminished or absent in all patients. Facial weakness ocurred in 4. CSF protein was elevated (mean, 177mg/dL) in 92%, but cells were not increased. Electrodiagnostic studies showed F-wave abnormalities (92%) and slowing of nerve conduction velocities (77%). Nerve biopsies performed in 4 showed demyelination. Prednisone resulted in initial improvement in all 13 patients. Relapses required continued prednisone in 8, and other therapies, such as immunoglobulin, plasma exchange, or immunosuppressive medications, were added. One group of patients (4) with weakness developing over a short period of 1 to 3 months showed a monophasic course with complete recovery in 3. A second group (9), with slower evolution of symptoms from 3 months to several years, had no complete recoveries and mild to severe residual weakness. (Nevo Y, Pestronk A et al. Childhood chronic inflammatory demyelinating neuropathies: clinical course and long-term follow-up. Neurology July 1996;47:98-102). (Respond: Dr Pestronk, Department of Neurology, Box 8111, 660 South Euclid Ave, St Louis, MO 63110).

COMMENT. Childhood onset chronic inflammatory demyelinating polyneuropathy (CIDP) has in general a poor long-term prognosis, the

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The editor is Pediatric Neurologist at Children's Memorial Hospital and Northwestern University Medical School, Chicago, Illinois.
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majority showing relapses and having residual weakness. After an initial improvement with prednisone therapy, attempts to withdraw steroids were often unsuccessful and the addition of immunosuppressive medications was rarely of benefit. The few children who recovered completely had an antecedent illness of URI or tonsillitis. Of two patients with CIDP associated with MMR immunization, none recovered and one had severe residual weakness. CIDP is a previously unreported side effect of MMR immunization. Transverse myelitis following MMR vaccine was reviewed in Ped Neur Briefs Sept 1995;9:65.

CONGENITAL AND JUVENILE MYASTHENIA GRAVIS

The clinical features, course, and presence of acetylcholine receptor antibody (AChRAb) were reviewed in 25 congenital (CMG) and 30 juvenile (JMG) cases of myasthenia gravis seen at Hacettepe University, Department of Paediatric Neurology, Ankara, Turkey. The age range of onset showed overlap: birth to 4 years for CMG, and 1.5 to 15 years for JMG. Parental consanguinity was present in 15 (60%) of CMG and only 3 (30%) of JMG patients. Motor development was delayed in 9 (36%) CMG infants and in 3 (10%) JMG patients. Initial symptoms were ocular in equal frequency for CMG (44%) and JMG (53%). After 1 year follow-up, only 4 (16%) CMG patients had ocular only involvement, 19 (76%) having progressed to ocular and bulbar or generalized weakness. Symptoms were limited to ocular muscles in 47% of JMG patients after 1 year. Unlike CMG, JMG patients showed spontaneous remissions in 20% and myasthenic crises in 33%. Good response to anticholinesterase drugs was more frequent in JMG than CMG (63 versus 41%). AChRAbs were present in 9 (34%) of JMG patients, all were girls with a later disease onset (>11 yrs) than antibody-negative cases. Pure ocular forms of MG were more often seronegative. None of the antibody-positive cases were in remission. The response to treatment was not significantly different between seropositive and negative cases. (Anlar B et al. Myasthenia gravis in childhood. Acta Paediatr July 1996;85:838-842). (Respond: Dr B Anlar, Hacettepe University, Department of Paediatric Neurology, Ankara 06100, Turkey).

COMMENT. In this series of childhood onset myasthenic patients, the proportion of congenital cases was much larger than previously reported. Facial muscle involvement and malformation often described in congenital cases was not alluded to in the above report. (see Progress in Pediatric Neurology I and II, PNB Publ, 1991 & 1994). Family and developmental histories, severity and distribution of weakness, and response to therapy are supportive criteria for the differentiation of congenital and juvenile cases.

GENETICS OF FACIOSCAPULOHUMERAL DYSTROPHY

The relationship of phenotype to genotype in a clinically and genetically well defined population of 157 affected patients and 62 kindreds with facioscapulohumeral muscular dystrophy (FSHD) was examined at the University of Rochester School of Medicine, NY, and Ohio State University, Columbus, OH. Using isometric myometry scores to quantify disease severity, a significant correlation between disease severity and the size of the 4q35-associated deletion was evident, and the offspring were more severely affected than their parents. This generation effect and presence of anticipation in FSHD suggests a possible underlying dynamic mutation and an unstable repeat element within the region of the 4q35 deletion. (Tawil R et al. Evidence for anticipation and association of deletion size with severity in