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## NEUROMUSCULAR DISORDERS

### IMMUNOGLOBULIN FOR POLYNEUROPATHY

The value of high-dose intravenous immunoglobulin in the treatment of chronic inflammatory demyelinating polyneuropathy was studied in a double-blind, placebo controlled, crossover investigation at the Department of Neurology, University Hospital Rotterdam-Dijkzigt, the Department of Immunohematology and Bloodbank, University Hospital Leiden, and the Central Laboratory of the Netherlands, Red Cross Blood Transfusion Service, Amsterdam, The Netherlands. Of seven patients treated two were children aged 10 and 7 and five were adults. At initial diagnosis one child was severely disabled and the other moderately disabled (Rankin score 5 and 3, respectively). All patients had weakness of both legs, areflexia, slowed nerve conduction velocities, and elevated CSF protein level. All had responded to treatment with IVIg, 0.4 g/kg bodyweight/2 weeks. All patients showed deterioration after IVIg was discontinued. The patients were then randomized to IVIg or placebo (albumin treatment) in a double-blind crossover study. Those treated with IVIg improved by day eight after the onset of treatment whereas those treated with placebo showed no improvement. The time lapse between discontinuation of the IVIg treatment until deterioration was 6.4 weeks. After placebo the time lapse in weeks until clinical deterioration was 1.3 weeks. (van Doorn PA et al. High-dose intravenous immunoglobulin treatment in chronic inflammatory demyelinating polyneuropathy: A double-blind, placebo-controlled, crossover study. Neurology Feb 1990; 40:209-212).

COMMENT. Chronic inflammatory demyelinating polyneuropathy may fluctuate in severity or show deterioration over many months or years and the course and prognosis differs from Guillain-Barre

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syndrome. Improvement has followed treatment with prednisone, plasmapheresis, and IV gamma globulin (Cook JD et al. Neurology 1987; 37 (suppl 1):253). High dose intravenous immunoglobulin is more convenient than plasma exchange and has less long-term side effects than corticosteroids. It was also of value in the treatment of two patients with demyelinating neuropathy associated with monoclonal gammopathy. (Cook D et al. Neurology Feb 1990; 40:212). Dyck PJ in an editorial comments that the processing of Ig is sufficiently rigorous that HIV or hepatitis should not be transmitted by IVIg and there are no other known complications from this treatment. The major drawback was the high cost, $750-1000/treatment.

SPINAL MUSCULAR ATROPHY

Juvenile chronic segmental spinal muscular atrophy of Hirayama is described in two adult identical twins from the Department of Neurology, University of Vermont College of Medicine, Burlington, VT. In both patients examined at 69 years of age, the weakness was first noticed in the right hand at age 16 and within six months there was similar weakness of the left hand and atrophy of muscles in both hands. The disability progressed over the next four years but after age 21, further decline was barely noticeable. Examination at age 69 showed marked atrophy and weakness of the intrinsic muscles of both hands, the ulnar half of the forearm flexors, and of the brachioradialis muscles more on the right side. Occasional fasciculations were noted in involved muscles. The triceps and Achilles reflexes were decreased in one patient and the tendon reflexes were otherwise normal. Nerve conduction studies showed reduced amplitude of the ulnar and median compound muscle action potential, mildly slow conduction in upper and lower extremity motor nerves, and mild prolongation of the F-wave latencies. EMG showed no fibrillations, positive waves or fasciculations. The motor units in the upper extremity muscles showed neurogenic features. Based on the identical sex, phenotypes, blood groups, and HLA typing in the two brothers there was a 98.8% calculated chance that these twins were identical. (Tandan R et al. Chronic segmental spinal muscular atrophy of upper extremities in identical twins. Neurology Feb 1990; 40:236-239).

COMMENT. Hirayama, the first to describe this disease, reported 38 cases and one autopsy report, showing loss of motor neurons and astrogliosis in C7/C8 anterior horns. A genetic cause for the disease is supported by its occurrence in identical twins.

CONGENITAL MYOPATHY IN LOWE SYNDROME

Congenital fiber type disproportion myopathy is described in two brothers with oculo-cerebro-renal syndrome of Lowe from the Department of Pediatrics, Tsuchiura Kyoudou Hospital; Ibaraki; Tsukuba University; Tokyo Medical and Dental University; and National Institute of Neuroscience; Kodaira, Japan. Both brothers had congenital cataracts, they were floppy as infants and psychomotor development was delayed.