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.

Both suffered febrile tonic seizures and one was hyperkinetic and had stereotypic hand movements. There was generalized muscle hypotonia, deep tendon reflexes were absent and serum creatine kinase activity was elevated. There was nystagmus in all directions of gaze. Biopsies of the biceps and brachia muscles showed selective type I fiber atrophy and mild type I fiber predominance similar to that observed in congenital fiber type disproportion myopathy. (Kohyama J et al. Congenital fiber type disproportion myopathy in Lowe syndrome. Pediatr Neurol Nov-Dec 1989; 5:373-376).

COMMENT. Selective type I muscle atrophy is uncommon and occurs in congenital nonprogressive myopathies, including nemaline myopathy, myotubular myopathy, central cord disease, and congenital fiber type disproportion. In association with type I fiber predominance it is observed in Pompe disease, Krabbe leukodystrophy and multiple sulfatase deficiency in which peripheral nerves are involved. The oculo-cerebro-renal syndrome of Lowe is characterized by mental retardation, glaucoma, congenital cataracts, and renal impairment. The elevated CPK led to the muscle biopsies in the present patients.

INTRACRANIAL TUMORS

HYPOTHALAMIC HAMARTOMA AND SEXUAL PRECOCITY

Four boys with hypothalamic hamartomas associated with sexual precocity are reported from the Departments of Pediatrics and Neurosurgery, University of Pittsburgh, and the Department of Pediatrics, Johns Hopkins University School of Medicine, Baltimore, MD. Two patients were treated surgically by resection using current microsurgical techniques and two received medical management. Precocious puberty caused by hamartomas occurs early in life with enlarged penis and muscular build noted in early infancy. Growth is accelerated and bone age is advanced. Deepening of the voice and the appearance of acne are common. Other clinical findings include mental retardation, behavioral disturbances, and seizures of gelastic, absence, and generalized tonic clonic patterns. In the two patients treated surgically, subsequent growth and development were normal and in two who were diagnosed late in childhood and treated medically, the adult height was not particularly compromised. The authors recommend surgical resection if the hamartoma is pedunculated or in cases where the patient is young and would require years of parenteral medical treatment. The nontreatment option exists because there is no evidence that the tumors will grow or subsequently cause other problems. (Starceski PJ et al. Hypothalamic hamartomas and sexual precocity. Evaluation of treatment options. AJDC Feb 1990; 144:225-228).

COMMENT. The use of MRI with sagittal, coronal, and axial views enable better visualization of hamartomas and earlier diagnosis. Treatment to allow growth and pubertal development to occur at an age appropriate time may avoid considerable