18 years, following the addition of carbamazepine, seizures were controlled, and pyridoxine was decreased to 100 mg daily. MRI showed left mesial temporal sclerosis. Nerve conduction studies at 2 years revealed absent sensory action potentials and normal motor conduction. Sural nerve biopsy showed severe, axonal, sensory neuropathy. At 18 years, vibration sense in the feet was absent, position sense was decreased in the toes, pain sensation was impaired to the midcalf and in the fingers, tendon reflexes were absent, and plantar responses were flexor. His gait was ataxic. Sural, peroneal and median sensory nerve action potentials were absent. The sensory neuronopathy diagnosed at 2 years had not progressed or remitted at 18 years. (McLachlan RS, Brown WF. Pyridoxine dependent epilepsy with iatrogenic sensory neuronopathy. Can J Neurol Sci February 1995;22:50-51). (Reprints: Dr RS McLachlan, University Hospital, 339 Windermere Rd, London, Ontario N6A 5A5).

> COMMENT. The authors explain the failure of pyridoxine to completely control the seizures in this patient by a combination of pyridoxine-dependent epilepsy with complex partial seizures due to mesial sclerosis. Unusually high doses of pyridoxine were prescribed in this patient. Doses as low as 50 mg/day have caused neuropathy when continued for months or years. Individual susceptibility is also a factor in the occurrence of this side effect.

CRITICAL ILLNESS NEUROMUSCULAR DISEASE

Four children with critical illness neuromuscular disease following prolonged dependency on a ventilator are reported from the Departments of Neurology and Pediatrics, West Virginia University Health Sciences Center, Morgantown, and the Department of Medicine (Neurology), Memorial University of Newfoundland, St John's, Canada. One patient, a 15-year-old boy with septic shock, required ventilatory support and intermittent vecuronium for neuromuscular blockade. Extubation on day 8 was unsuccessful because of quadriparesis, with diffuse muscle atrophy, and absent reflexes. Muscle strength gradually returned over 3 months, but hyporeflexia persisted for > 1 year. (Sheth RD, Bodensteiner JB et al. Critical illness neuromuscular disease in children manifested as ventilatory dependence. J Pediatr February 1995;126:259-61). (Reprints: Raj D Sheth MD, West Virginia University Health Science Center, Box 9180, Morgantown, WV 26506).

> COMMENT. Critical-illness polyneuropathy, a complication of sepsis in adults, and a cause of difficulty in weaning from the ventilator, is covered in Progress in Pediatric Neurology II, 1994, pp275-276. The syndrome appears to be unusual in children.

MUSCULAR FATIGUE IN DUCHENNE DYSTROPHY

The fatigability of the anterior tibial muscle in 11 boys with Duchenne muscular dystrophy (DMD) was compared to that of controls at the California Pacific Medical Center and the University of California, San Francisco. The force generation of dystrophic muscle and compound muscle action potential amplitude were lower and relaxation time of tetanus was longer in patients than in controls at rest. During exercise, maximum voluntary contraction was better sustained, suggesting less central fatigue in DMD patients than in controls. (Sharma KR, Mynhier MA, Miller RG. Muscular fatigue in Duchenne muscular dystrophy. Neurology February 1995;45:306-310). (Reprints: Dr Khema R Sharma, University of Miami, Dept of Neurology, 1501 NW 9th Ave, Miami, FL 33136).

COMMENT. The intramuscular fatigability and recovery following sustained maximum voluntary contraction was similar in dystrophic muscles and controls, but patients with DMD had less central fatigue, possibly explained by longer training sessions and greater familiarity with the exercise.

PELIZAEUS-MERZBACHER DISEASE AND HYPOTONIA

Two brothers with Pelizaeus-Merzbacher disease presenting with neonatal hypotonia and hyporeflexia are reported from the Tuft's University School of Medicine, and Massachusetts General Hospital, Boston, and the EKS Center for Mental Retardation, Waltham, MA. Patient 1 had Apgar scores of 0 at 1 minute and 3 at 5 minutes. The neonatal exam showed signs of spinal muscular atrophy: poor suck and swallow, tongue fasciculations, reduced muscle bulk and strength, severe hypotonia, absent deep tendon reflexes, and normal sensation. An older brother had a similar disease. Parents were not related. Nystagmoid eye movements were present shortly after birth and disappeared before 1 year. Optic atrophy and dystonic arm movements developed after several months. MRI showed diffuse hypomyelination. EMG demonstrated fibrillations, high-amplitude potentials, and acute denervation. Muscle biopsy was normal. The diagnosis of Pelizaeus-Merzbacher disease was confirmed by proteolipid protein genetic studies. (Kaye EM et al. Pelizaeus-Merzbacher disease presenting as spinal muscular atrophy: Clinical and molecular studies. Ann Neurol Dec 1994;36:916-919). (Respond: Dr Kaye, Division of Pediatric Neurology, Floating Hospital for Children, Tuft's University School of Medicine, Boston, MA 02111).

COMMENT. Pelizaeus-Merzbacher disease should be included in the differential diagnosis of limp infant syndrome and as a cause of infantile spinal muscular atrophy.

NEUROCUTANEOUS SYNDROMES

LEUKOENCEPHALOPATHY AND KERATODERMA

A new familial neurocutaneous syndrome consisting of palmoplantar keratoderma (PPK) and adult-onset leukoencephalopathy is reported in four siblings from Hadassah University Hospital, Jerusalem, Israel. The proband, a 41-year-old woman, had a progressive gait disorder and cognitive impairment of 3 years' duration. Skin abnormalities had been present from early childhood: thick hyperkeratotic skin with numerous papules, most prominent over the palmar and plantar areas. Neurologic exam revealed cognitive impairments, generalized weakness with symmetric hyperreflexia, hypertonia, and Babinski signs. CSF protein was 1.18 g/L. MRI showed periventricular hyperintensity of white matter, brain atrophy, and thinning of the corpus callosum. Arylsulfatase A pseudodeficiency carrier state was identified by molecular analysis. Sural nerve biopsy showed loss of myelinated fibers. Bone X-rays showed osteochondritis dissecans of the talus. (Lossos A et al. Hereditary leukoencephalopathy and palmoplantar keratoderma: A new disorder with increased skin collagen content. Neurology Feb 1995;45:331-337). (Reprints: Dr A Lossos, Dept Neurol, Hadassah Univ Hosp, Jerusalem 91120, Israel.

COMMENT. Neurologic complications of PPK are rare and late in onset.