tetraparesis with cerebellar signs, but without spasticity and signs of pyramidal dysfunction. The disease has greater clinical vartiability than previously recognized. MRI findings can be helpful in diagnosis, with early involvement and atrophy of the cerebellar vermis. The pallidal involvement in 2 of the above series suggests an overlap with the radiologic findings in Hallervorden-Spatz disease. A biochemical or molecular marker has not been uncovered.

NEUROMUSCULAR DISEASES

OUTCOME IN SEVERE GUILLAIN-BARRE SYNDROME

The effect of various therapies on duration of illness in children with severe Guillain-Barre syndrome (GBS) was evaluated at CHMC, Seattle, WA. Of 26 children treated in two contiguous 8-year periods, 12 received supportive care alone (SC), and 14 were treated with SC plus either plasma exchange (PE), 6 cases, or intravenous immunoglobulin (IVIg), 8 cases. Recovery to score 2 (able to walk 5 m without support) was similar in the SC and IVIg groups, with a nonsignificant trend toward longer recovery times in the PE group. The addition of PE or IVIg did not improve outcome or shorten duration of illness compared with SC alone. Full recovery occurred in almost all patients within 6 months of disease onset. (Graf WD, Katz JS, Eder DN, Smith AJ, Chun MR. Outcome in severe Guillain-Barre syndrome after immunotherapy or supportive care. Neurology April 1999;52:1494-1497). (Respond: WD Graf MD, Division of Child Neurology, CHMC, 4800 Sandpoint Way, Seattle, WA 98105).

COMMENT. Immunotherapy in severe pediatric GBS does not improve outcome or shorten duration of illness compared with supportive care alone and may be less effective than in adult cases of GBS.

BETHLEM MYOPATHY

The natural course of Bethlem myopathy in five previously published kindreds and two novel pedigrees was investigated, with attention to the mode of onset in 23 children and the progression of weakness in 36 adult patients followed at the Academic Medical Center, Amsterdam, The Netherlands. Onset was characterized by diminished fetal movements, neonatal hypotonia and congenital contractures including torticollis, nearly all children exhibiting weakness or contractures and slightly delayed milestones during the first 2 years of life. Symptoms became more evident at 5 years of age, with worsening of contractures and weakness during childhood, followed by relative recovery during puberty. During early adult life, many patients were nearly asymptomatic except for contractures. From middle age onwards, the contractures remained constant but weakness and incapacity showed slow but ongoing progression into adulthood, more than two-thirds of patients over 50 years of age requiring a wheelchair. (Jobsis GJ, Boers JM, Barth PG, de Visser M. Bethlem myopathy: a slowly progressive congenital muscular dystrophy with contractures. Brain April 1999;122:649-655). (Respond: Dr GJ Jobsis, Department of Neurology, (H2-214), Academic Medical Center, PO Box 22700, 1100 DE Amsterdam, The Netherlands).

COMMENT. In 1976, Bethlem and van Wijngaarden described three families with an early-onset benign autosomal dominant myopathy with contractures. Contractures involved fingers, wrists, elbows, shoulders, knees and hips. Weakness was mild, affecting proximal and extensor muscles, with only limited functional impairment, even in old age. Cranial and cardiac muscles were not