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

involved. Serum CPK was normal or slightly elevated and muscle biopsy changes were nonspecific. Subsequent reports included slightly variable manifestations, some children showing highly elevated CPK, up to 15 times normal, and muscle biopsies with dystrophic, necrotic and degenerating changes. Genetic tests find collagen type VI as the defective protein in Bethlem myopathy. The present study defines further variations in the clinical manifestations of the disease, with onset of symptoms at or even before birth, and a slowly progressive course in middle to late adulthood.

ATTENTION DEFICIT DISORDER

METHYLPHENIDATE AND CLONIDINE COMBINATION DEBATED

The Debate Forum of the J Am Acad Child Adolesc Psychiatry, in the May 1999 issue, addresses the concern regarding reported risk of fatalities in children receiving clonidine in combination with methylphenidate (MPH) for the treatment of ADHD with comorbid aggression, Tourette syndrome, or sleep disturbance.

Swanson JM, Connor DF, and Cantwell D consider the combination ill-advised, pending the results of controlled studies at the NIMH and the University of Massachusetts, and citing "the absence of clear demonstration of efficacy, and in the face of plausible but not-yet-validated serious side effects of the combination."

Wilens TE and Spencer TJ support the use of this combined therapy as a clinically sound medication option, citing promise in open trials and growing popularity with "widespread clinical use." These authors find the evidence for clonidine/MPH fatalities in 4 reported cases weakened by complicating factors, including congenital cardiac malformation. They do agree, however, on the need for further monitoring and study of the safety of the combination.

Swanson and associates, in a further negative rebuttal, respectfully disagree with the acceptance of uncontrolled trials as evidence for cited efficacy of the clonidine/MPH combination. They emphasize the relatively high (5%) drug-related electrocardiographic irregularities reported with clonidine, and the dangers of rising and falling levels of clonidine and MPH and their effects on cardiac function. (Debate Forum. Combining methylphenidate and clonidine. J Am Acad Child Adolesc Psychiatry May 1999;38:614-622).

COMMENT. This exchange of views regarding the use of combined methylphenidate and clonidine therapies for ADHD with comorbid aggression, Tourette syndrome, or sleep disturbance is very informative, but the opposing opinions of experts may leave the practicing neurologist or psychiatrist in a dilemma when faced with the decision to risk a possible fatality as a result of prescribed treatment. I tend to agree with the Swanson team that drug combinations with reported serious, and sometimes fatal, cardiac side effects should be withheld pending definitive controlled studies proving safety and efficacy. (see Millichap JG. Attention Deficit Hyperactivity and Learning Disorders. Chicago, PNB Publishers, 1998;p187-9).

Methylphenidate and behavior modification had beneficial effects on ADHD symptoms and comorbid ODD in a randomized, placebo-controlled study of 16 children studied at the Western Psychiatric Institute and Clinic, University of Pittsburgh Medical Center (Kolko DJ, Bukstein OG, Barron J. J Am Acad Child Adolesc Psychiatry May 1999;38:578-586).