

patients with other forms of muscular dystrophy and in control subjects.

DUCHENNE MUSCULAR DYSTROPHY

The clinical progression and effects of therapy in 283 boys with Duchenne dystrophy and ten with Becker dystrophy followed for up to ten years in a collaborative study are reported from the Departments of Neurology and Biostatistics, Washington University School of Medicine, St. Louis, MO, the Departments of Neurology, Vanderbilt University, Nashville, TN, Ohio State University, Columbus, Ohio and University of Rochester, Rochester, New York. The protocol measured function, strength, contractures, and scoliosis. A series of milestones allowed the severity of the disease to be defined in an individual boy. After age 11, 89 of 120 patients developed a scoliosis. The use of a body jacket to control a progressive scoliosis was ineffective but back surgery was beneficial if carried out before the forced vital capacity was less than 1.5 liters. The average age at the time of surgery was 14.6 years and patients with a curve exceeding 35° were considered to be candidates for surgery. No correlation could be detected between the use of passive joint stretching exercises and joint contractures but there was a significant correlation between the use of leg braces and the prevention of contractures of the heel cords, knee extensors, and iliotal bands. There were 25 deaths while the boys were enrolled in the protocol. Most deaths occurred from respiratory failure, often after repetitive bouts of pneumonia, or from cardiac failure. Weaker patients died from respiratory failure whereas those whose muscles were stronger were more likely to die from a cardiomyopathy. (Brooke MH, Fenichel GM et al. Duchenne muscular dystrophy: Patterns of clinical progression and effects of supportive therapy. Neurology April 1989; 39:475-481).

COMMENT. In the same issue of Neurology, genetic abnormalities in Duchenne and Becker dystrophies with clinical correlations (Medori R et al) and molecular and clinical correlations of deletions leading to Duchenne and Becker muscular dystrophies (Baumbach LL et al) are reported. Duchenne muscular dystrophy (DMD) and Becker muscular dystrophy (BMD) are two allelic forms of an x-linked muscle disorder with phenotypic heterogeneity. Of 32 DMD patients, 14 had an internal deletion in the same region of the gene and 7 of 11 patients with a mild DMD or BMD phenotype showed deletions at the 5' end of the gene. Patients with classic DMD who had a detectable deletion had a milder clinical course than those without. BMD patients may be genetically different from boys with classic DMD. There was no correlation between the extent of a deletion, its location, and clinical severity of the associated disease. Duchenne muscular dystrophy is a severe x-linked disease with an incidence of 1 in 3500 males; approximately one-third result from a new mutation. Becker muscular dystrophy is a clinically similar but less severe form of dystrophy affecting 1 in 30,000 males. The application of recombinant DNA technology to the diagnosis of DMD has resulted in the development of more accurate tests which supplement the serum CPK, muscle biopsy and EMG.