normal control group. The mean area of the superior-posterior vermis in the autistic subject group was 20% smaller than in the normal control group, while there was no significant difference between the mean anterior vermis areas of the two groups. The results indicated that the decreased size of the cerebellar hemispheres and the vermal lobules VI through VII was associated with autism. (Murakami JW et al. Reduced cerebellar hemisphere size and its relationship to vermal hypoplasia in autism. Arch Neurol June 1989; 46:689-694).

COMMENT. The results of this study confirm those of a previous study by the same authors that showed that hypoplasia of the superior-posterior vermis (lobules VI and VII) is frequently observed in autistic individuals. The nature of the link between cerebellar dysgenesis and autistic symptoms has not been determined. The authors refer to clinical and research observations indicating that the cerebellum also plays a role in a variety of cognitive functions, such as language, learning and memory, emotional behavior, and complex motivated behaviors. They believe that the hypoplasia of cerebellar hemispheres and vermis observed in many autistic individuals is linked with behavioral and cognitive symptoms.

MUSCLE DISORDERS

SELENIUM AND MUSCULAR DYSTROPHY

Selenium metabolism and supplementation in patients with Duchenne muscular dystrophy was studied at the Muscle Research Center, Department of Medicine, University of Liverpool, and the Universitat Klinik Mainz, Mainz, FRG. Plasma selenium concentrations measured in seven Duchenne muscular dystrophy patients and in 11 age matched normal boys showed no significant difference after two months of sodium selenite supplementation (1 mg selenium daily). All patients demonstrated a rise in plasma selenium concentration as did all but one of the normal subjects. The studies did not confirm any abnormality of selenium metabolism in patients with muscular dystrophies, and there was no evidence that high dose selenium supplementation influenced the activity of the selenium dependent enzyme glutathione peroxidase in skeletal muscle. An elevation of thiobarbituric acid-reacting substances in the muscle of patients with Duchenne muscular dystrophy was unaffected by selenium supplementation (Jackson MJ et al. Selenium metabolism and supplementation in patients with muscular dystrophy. Neurology May 1989; 39:655-659).

COMMENT. The present finding of normal plasma selenium concentrations in Duchenne muscular dystrophy patients differs from reports from Finland where selenium in soils and indigenous food stuffs is naturally low in concentration. The increase in thiobarbituric acid-reacting substances in dystrophic muscle confirms previous reports but the elevated levels in patients with Duchenne muscular dystrophy contrasted with normal levels in

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 iliotibial 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, Fenischel 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.