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MUSCLE DISORDERS

LANGUAGE DELAY IN DUCHENNE'S MUSCULAR DYSTROPHY

Developmental milestones of 130 male children with Duchenne's muscular dystrophy (DMD) and their 59 unaffected siblings were determined by retrospective parental reports and compared by researchers at the City University and Columbia University, New York, NY; and Scottish Rite Children's Medical Center, Atlanta, GA. Participants were recruited through the Muscular Dystrophy Association clinics at these institutions. Children with DMD were 4-14 years of age (mean, 9 years), and sibling controls 3-16 years (mean, 9.85 years). Parents completed a developmental milestone questionnaire and Child Behavior Checklist. Neuropsychological testing included measures of language, memory, and visuospatial skills (Peabody Picture Vocabulary Test, and the Raven's Colored Progressive Matrices). Children with DMD were rated late more often than their unaffected siblings in motor milestones such as sitting (38% vs 0%), crawling (60% vs 6%), standing (56% vs 0%), and walking (70% vs 2%)($P<.001$). They were also late in speaking their first word (42% vs 4%) and in using full sentences (49% vs 4%)($P<.001$), and in reading (94% vs 6%)($P<.001$). Development of other milestones (smiling, bowel or bladder control) was not delayed. DMD children late in constructing complete sentences performed poorly on measures of single-word vocabulary ($P<.001$) and visuospatial reasoning ($P=.002$), compared to DMD children whose language development was on-time. Behavioral difficulties were not significantly different in the two groups of DMD children. DMD children rated late in walking performed more poorly on a measure of visuospatial reasoning ($P<.02$), but were not different in acquiring single-word vocabulary nor in frequency of behavioral issues when compared with DMD early walkers. (Cyrulnik SE, Fee RJ, De Vivo DC et al. Delayed developmental language milestones in children with Duchenne's muscular dystrophy. *J Pediatr* May 2007;150:474-478). (Respond: Veronica J Hinton PhD, Columbia U, NY).

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COMMENT. In addition to delays in motor milestones, children with DMD are late in speaking single words and sentences. The late talkers also develop cognitive deficits involving vocabulary. Both late talkers and late walkers function poorly on tests of visuospatial reasoning. Age at taking the first step is generally considered the most reliable milestone based on parents' recollection, while sitting and uttering the first word are less well timed. In the present study, by focusing on broad categories ("on-time" and "late") and by using siblings as controls, the authors have attempted to minimize the limitations of a retrospective study.

Although DMD is primarily a disease of muscle, unexplained language and developmental delay may precede significant signs of muscle weakness, and point to a coincident CNS disorder. Deficiency of brain synaptic dystrophin was demonstrated in an 8-year-old child with DMD examined at autopsy (Kim T-W et al. *Ann Neurol* 1995;38:446-449). Several studies have demonstrated the association of cognitive deficits, especially specific verbal impairments in children and adolescents with DMD. The present report shows that delayed language milestones may be the earliest sign of DMD, and an important observation in the clinical diagnosis. The need for further study of brain dysfunction in children with DMD is indicated.

OUTCOME OF LONG-TERM CORTICOSTEROID TREATMENT IN DUCHENNE MUSCULAR DYSTROPHY

The clinical orthopedic effects of chronic daily corticosteroid treatment were evaluated by chart review in boys with genetically confirmed Duchenne muscular dystrophy (DMD) followed at the Ohio State University Muscular Dystrophy Clinic between 2000 and 2003. Becker dystrophy cases diagnosed in 16 were excluded. Of 143 with DMD, 75 (52.4%) had been treated with steroids for at least 1 year (prednisone 0.75 mg/kg/day or deflazacort 0.9 mg/kg/day), and 68 (47.6%) had never been treated or had received brief trials of subtherapeutic doses (5 mg prednisone daily for < 6 months). Both cohorts had a mean height below the 5th percentile. Mean age of the treated group was 16.9 years (range 6.1-30.5 years) vs 14.4 (1.1-39.6 years) for untreated patients. Reason for non-treatment was invariably parent fear of side effects. Mean duration of steroid treatment was 8.04 years (range 1-18 years). Independent ambulation was lost at 12.52 +/- 3.02 years for steroid-treated vs 9.21 +/- 1.48 years in untreated patients (P<.0001). Lower limb fractures accounted for loss of ambulation in only 1 case (3.1%) in the untreated group compared to 11 (40.7%) of the steroid-treated patients. Scoliosis of >10 degrees developed in 31% of treated vs 91% of untreated (P<.0001). Vertebral compression fractures occurred in 32% of steroid-treated and none of untreated patients (P=.0012). The prevalence of femoral fractures was 28.3% in treated vs 7.27% in untreated patients (P=.0051). Humeral fractures were less frequent in steroid-treated (9.43%) compared to untreated patients (25.45%) (P=.042). (King WM, Ruttencutter R, Nagaraja HN et al. Orthopedic outcomes of long-term daily corticosteroid treatment in Duchenne muscular dystrophy. *Neurology* May 2007;68:1607-1613). (Reprints: Dr JT Kissel, Department of Neurology, Division of Neuromuscular Disease, Ohio State University Medical Center, 1654 Upham Dr, Columbus, OH 43210).