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SPINAL DEFORMITY IN PROGRESSIVE NEUROMUSCULAR DISEASE

Natural History and Management

Dennis A. Hart, MD, and Craig M. McDonald, MD

Severe spinal deformity in progressive neuromuscular disease (NMD) leads to multiple problems, including poor sitting balance, difficulty with upright seating and positioning, pain, difficulty in attendant care, and potential exacerbation of underlying restrictive respiratory compromise. Severe scoliosis and pelvic obliquity can in some instances completely preclude upright sitting in a wheelchair. Populations at risk for scoliosis include Duchenne muscular dystrophy (DMD), severe childhood autosomal recessive muscular dystrophy (SCARMD), congenital muscular dystrophy (C-MD), facioscapulohumeral muscular dystrophv (FSH), congenital myotonic muscular dystrophy, spinal muscular at phy II and III, and Friedreich's ataxia. This article reviews the pre lence, natural history, and management of spinal deformity in th progressive neuromuscular disorders.

DUCHENNE MUSCULAR DYSTROPHY

Prevalence and Natural History

The reported ultimate prevalence of scoliosis in DMD varies fr 33% to 100%.[18, 26, 31, 41, 44, 50, 60, 61, 70, 71, 74, 77, 80, 89] This marked variabilit

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Prevalence and Natural History

The reported ultimate prevalence of scoliosis in DMD varies from 33% to 100%.[18, 26, 31, 41, 44, 50, 60, 61, 70, 71, 74, 77, 80, 89] This marked variability is

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primarily because of retrospective selection for scoliosis, the inclusion or exclusion of functional curves, and dissimilar age groups. As shown in several studies,[50, 52, 60] the prevalence of scoliosis is strongly related to age. Fifty percent of patients acquire scoliosis between ages 12 and 15, corresponding to the adolescent growth spurt. Figure 1*A* reflects the relationship between age, adolescent growth spurt, and onset of scoliosis. Note that 10% of older DMD subjects in McDonald's series[52] show no clinical scoliosis. This is consistent with Oda's report[60] of a 15% prevalence of DMD patients with mild nonprogressive curves (usually 10° to 30°). The rate of progression of the primary or single untreated

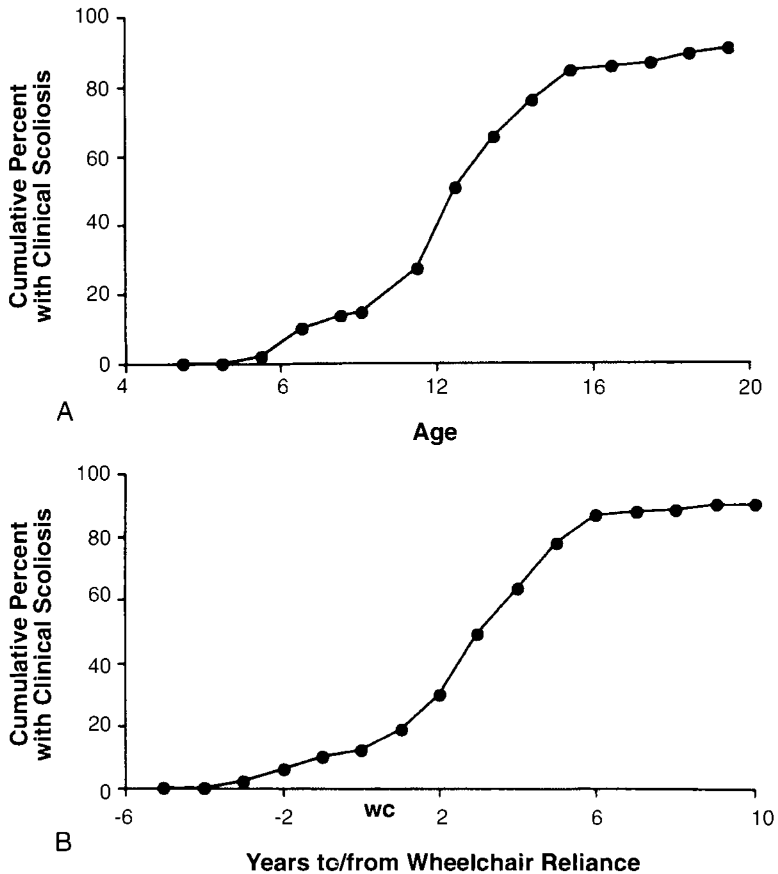

Figure 1. Cumulative percentages of Duchenne muscular dystrophy (DMD) subjects with scoliosis on clinical examination *v (A)* age and *(B)* years to and from consistent wheelchair reliance. (*From* McDonald C, Abresch T, Carter G, et al: Profiles of neuromuscular diseases: Duchenne muscular dystrophy. Am J Phys Med Rehabil 74(supp 5):S70–S92, 1995; with permission.)

lateral curve has been reported to range from 11° to 42° per year, depending on the age span studied.

Johnson and Yarnell[44] report an association between side of curvature convexity and hand dominance. McDonald's study,[52] on the other hand, showed no correlation between side of primary convexity and handedness. Thus, placement of the power wheel chair drive control in the midline to decrease lateral trunk bending has never been documented to alter the natural history of spinal deformity in DMD.

Oda et al[60] reviewed the natural history of scoliosis in DMD. They classified the course of spinal deformity into three types (Fig. 2). Type 1 curves follow an unstable course and are characterized by progression of scoliosis concomitant with kyphosis. This progression leads to a collapsing spine with significant rotatory deformity. The lateral curvature presents in the thoracic or thoracolumbar region and involves the sacrum and pelvis. The relationship of lateral curvature to pelvic obliquity has been reported in other studies.[60, 80, 89] In type 1 spinal deformity the scoliosis always reaches greater than 30° before age 15, with rapid progression after 30°. Surgical stabilization is indicated after the curve reaches 30° and before vital capacity reaches 35% of predicted.

Type 2 curves are characterized by transition of kyphosis to hyperlordosis in the sagittal plane, associated with progression of scoliosis. The course of the scoliosis in type 2 patients is not uniform. Patients with a double lateral curve show little change in Cobb angle and have minimal pelvic obliquity. Patients with long thoracolumbar curves (Fig. 2) showed gradual increase in severity of deformity. In patients with single lumbar curves the Cobb angle may show gradual increase or sudden increase. As in type 1 curves, lumbar and long thoracolumbar type 2 curves are associated with pelvic obliquity. Oda's study could not definitively offer early predictive indicators of later required surgical instrumentation for type 2 patients.

Type 3 curves (Fig. 3) show minimal progression, with the Cobb angle never reaching greater than 30°. There is no indication for surgical instrumentation for type 3 curves in DMD. Oda's study also demonstrates a better prognosis for pulmonary function in type 3 spinal deformity, with peak obtained absolute functional vital capacity (FVC) greater than 2000 mL. Most type 1 and type 2 patients showed less than 2000 mL peak obtained FVC.

The Role of Wheelchair Reliance in the Pathogenesis of Spinal Deformity in DMD

No cause-and-effect relationship has been established between onset of wheelchair reliance and occurrence of scoliosis. However, in patients developing scoliosis, severity increases with age and length of time in a wheelchair.[50, 52] Figure 1*B* reflects the cumulative percentages of DMD with scoliosis versus years to and from wheelchair reliance. Note that approximately 15% develop scoliosis before transition to a wheelchair.