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Friedreich's ataxia is frequently classified as a distal motor and sensory neuropathy with a predominantly distal muscle weakness, similar to that in Charcot-Marie-Tooth disease. There is limited published data to support this statement. We have studied retrospectively 201 detailed and standardized muscle-strength evaluations done between 10 and 30 years (mean 20.4 years) and have correlated them with ambulatory status, using Hoffer's classification. All muscle testings evaluated the same 27 main muscle-groups, graded from 0 to 5, and were done by the same physiotherapist. Results show that the first muscle-group significantly weakened is the hip extensor in 85 per cent of patients, and that the pattern of muscle weakness is always symmetrical, initially proximal rather than distal, more severe in the lower extremities and rapidly progressive once the patients become non-ambulators. Over-all muscle strength in the lower extremities is 72 ± 6.1 per cent of normal when community ambulators become household ambulators, and 63.2 ± 8.5 per cent when functional ambulation is lost, thus suggesting that muscle weakness is not the primary cause of loss of ambulatory function.

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Quantification of Spasticity with the Torque-velocity Curve

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This study quantified the torque-velocity (T-V) relationship for the knee extensors of patients with cerebral palsy and of controls. It was hypothesized that the slope of the T-V regression line would be lower for patients than controls.

Thirty-two patients (64 limbs) with cerebral palsy were evaluated. Their mean age was $9.1 \pm$ four years. There were 25 control subjects (50 knees), whose mean

age was $10.1 \pm$ four years. Torque and angular displacement were measured with an isokinetic dynamometer, modified with a strain-gauge torque sensor and equipped with an electrogoniometer. Isokinetic torque values were normalized by calculating (isokinetic torque/isometric torque) $\times 100$ for each isokinetic speed. The T-V slopes were calculated from these normalized values.

The patients' T-V slope, -0.50 ± 0.22 Newton-meters/degree/second (N-m/d/s) was significantly lower ($p < 0.001$) than the slope for the controls, -0.27 ± 0.10 . In patients, the T-V slope was significantly lower for non-ambulators (-0.68 ± 0.25 N-m/d/s) than for ambulators (-0.40 ± 0.16 N-m/d/s). The ambulators had a trend toward higher slopes for those who ambulated at a faster preferred rate.

Torque production decreased more rapidly with increasing angular velocity in patients than in controls. This may be due to the spastic response of the antagonist, impairment of agonist control, or the complex interaction of both factors. The T-V slope is related to ambulatory status. Data will be presented on the relationship with the ordinal severity of spasticity. In addition, results will be presented on performing these evaluations before and after hamstring tenotomy.

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The Influence of Inclined Wedge Sitting on Infantile Postural Kyphosis

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The purpose of this research study was to determine whether the kyphotic sitting posture of developmentally delayed children could be influenced by altering the angle of the pelvis. There were 16 children, eight girls and eight boys. Their sitting postures were marked by postural kyphosis, posteriorly tilted pelvis, and lower extremity flexion, abduction and external rotation.

The children were placed in sitting with a grid to their right. Rubber darts were taped on and used as markers on the vertebrae: C7, T7, T12/L1 and S1. Each

child was videotaped for 60-second intervals during the following sequence: on the mat, on a 15° wedge, on the mat, on the 25° wedge, and on the mat. The data was collected by drawing vectors between the C7 and T7 markers, the T7 and T12/L1 markers, and the T12/L1 and S1 markers. The vectors, along with a relative vertical line, created an angle for the measurement of each respective section, referred to as the superior, middle and inferior segments.

The children demonstrated less postural kyphosis on the 15° and 25° wedges. On the 15° incline the outcome was statistically significant for all three segments. The changes on the 25° wedge proved to be statistically significant in the middle and inferior segments.

The results of this study suggest that the kyphotic sitting posture of developmentally delayed children may be improved by using a posteriorly inclined wedge for short periods of time. Biomechanical and neurophysiological rationale will be offered to support the outcome of this study.

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Hip Angle and Upper-extremity Movement Time of Children with Cerebral Palsy

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This study was designed to evaluate the effect of hip-flexion angle on upper-extremity function of patients with cerebral palsy.

The study included 10 children diagnosed as having cerebral palsy. They were placed in an adjustable seating device so that the head, neck and trunk were in the vertical plane, with the knee and ankle at 90°. The hip was then randomly placed at 50°, 70°, 90° and 110° of flexion. The children were instructed to perform horizontal adduction at each hip-flexion position to trigger a touch-activating switch on cue from a television monitor. 10 movements were performed and the average movement time (the time-interval between the cue to move the arm and activation of

the switch) was calculated for each hip-flexion position.

The mean upper-extremity movement time obtained with the hip at 50° was 1.87 seconds. Mean movement times of 1.52 seconds, 1.07 seconds and 1.48 seconds were obtained at 70°, 90° and 110° hip-positions, respectively. The results of analysis of variance indicated that a significant difference existed between the means at the 0.05 level. However, Fisher Multiple Comparisons showed that the significant difference was between the 90° position and the 50°, 70° and 110° positions. The difference between the 50° position and the 70° and 110° approached a significant level (0.06). The means at 70° and 110° were not significantly different.

For the type of patients and at the upright body-orientation used, the results of this study indicate that hip-flexion angle affects upper-extremity movement time. At the 50° position it was observed that breathing was much more labored, perhaps due to a decrease in the thoracic and abdominal cavities, and may have affected upper-extremity movement.

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Effect of Adaptive Seating on Pulmonary Function of Children with Cerebral Palsy

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This study was designed to measure the vital capacity (VC) and forced expiratory volume in one second (FEV₁) of children with cerebral palsy when seated in a regular sling-type wheelchair and in an adaptive seating system. The purpose was to determine the comparative effects of adaptive and non-adaptive seating on pulmonary function.

Nine children between the ages of five and 12 years diagnosed as having cerebral palsy were used for the study. VC and FEV₁ were obtained using a spirometer under two seating conditions. In one condition (non-adaptive seating) the child was seated in a regular, growing-size, sling-type wheelchair; in the other