

## XII. Muscular Dystrophy

### A Study of the Mechanism of Spinal Collapse in Duchenne Muscular Dystrophy \_\_\_\_\_

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**Purpose**—Increasing spinal deformity is a distressing feature of Duchenne muscular dystrophy. The orthotic nonsurgical management of paralytic spinal deformity has been written about extensively, with conflicting results concerning effectiveness. Our own work has indicated that external supports are not effective in keeping the spine straight and that lumbar pads are not effective in supporting the spine against lateral curvature. Still lacking is a mathematical model of the spine which would lead to a greater depth of understanding of the results we see in terms of the geometry and forces involved. Our goal is to develop a mathematical model, or models, of the spine and its muscles to answer these questions:

- 1) Can orthotic supports apply sufficiently high forces to straighten the spine without exceeding pad-to-skin pressures which the boys can tolerate?
- 2) Does a hyperextended lumbar spine, combined with rigid contracture of the paraspinal muscles, result in increased resistance to lateral bending? If so, what degree of hyperextension is required? Is this tolerable to the boys?
- 3) Is it true that supporting oneself on the nondominant arm creates a spinal curve with convexity towards the dominant hand?
- 4) What factors determine whether the curve is C-shaped or S-shaped?
- 5) Is a flat seat or a sling seat better? (Flat seats are often said to be better, but it may be that a sling seat allows the pelvis to tilt so that the head may be brought into line over the pelvis, and this may be preferable.)
- 6) In what way is lateral curvature related to rotation about the long axis of the spine? Is this related to the line of pull of muscles?
- 7) Why do the boys lean forward in their wheelchairs? Is this related to balancing the head on the spine by placing the neck in extension to relieve the neck extensors?
- 8) Is it possible to recommend any strategies for posture support in special seating as a result of this study.

**Progress**—*Preliminary phase:* Conduct a literature search to locate the models which may be most applicable to this study. Construct a simple model consisting of a slender beam and rigid support wires as a first model.

*Experimental phase:* Construct an accurate model appropriate to this study which represents the head, vertebrae, and spinal muscles.

*Concluding phase:* Publish the results of this study in a professional scientific journal. It is important that the results of this study are sufficiently convincing to influence the thinking of medical and allied health professionals regarding the treatment of paralytic scoliosis.

A mathematical model of the paralytic spine could lead to a greater understanding of the spinal collapse so often seen in Duchenne muscular

dystrophy. It would provide a theoretical underpinning to the spinal bracing and posture supporting seating provided at Regency Park Centre. This study should provide useful further information for medical and allied health professional who treat these children.

## Emotional Disorder Associated with Duchenne Muscular Dystrophy ---

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**Purpose**—The hypothesis of this study is that boys with Duchenne Muscular Dystrophy suffer from more depression and anxiety than a normal control group.

**Progress**—Twenty-three boys (4 to 17 years) were studied. Five of the boys who were mentally handicapped or of dull normal intelligence were excluded, leaving 18 boys in the study. These boys were matched for age, I.Q., and social class with a control group of boys without physical handicap attending normal schools. All boys and their parents were interviewed using the Rutter and Graham standardized psychiatric interviews for detection of psychiatric disorder in children. Psychological assessment of I.Q. was carried out, and teachers completed a Rutter B scale for each boy.

The prevalence of psychiatric disorder was significantly higher in the Muscular Dystrophy group than in the controls. Eight of the Muscular Dystrophy boys (44 percent) were diagnosed as having an emotional disorder of the mixed type with poor peer relationships, solitariness, anxiety, and depression as the main features. Older boys were at greater risk of psychiatric disorder than younger boys. Two of the controls (10 percent) were diagnosed as having an emotional disorder with anxiety. The study attempted to make a more precise diagnosis of affective disorder using DSM-III criteria, but as all the study boys had sleep problems, poor energy, and motor retardation as part of their Muscular Dystrophy this was felt not to be appropriate. Only two of the 18 boys had ever questioned their parents about their condition, and most parents felt unable to discuss the condition with their boys. Nevertheless, many families seemed to cope remarkably well with the condition using silence as part of their coping style.

**Future Plans**—Further investigation of this hypothesis will be conducted and the study will be extended to include a control group with a comparable physical handicap which is non-fatal (e.g. spina bifida).

## An Arm-Lifting Device ---

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**Sponsor:** The Northern Ireland  
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**Progress**—In response to the needs of children with Muscular Dystrophy, a device has been made that permits the arms to be elevated. It is a simple gantry attached to a wheelchair, that suspends the arms on plastic arm-rests. Normally, counterweights compensate for the weight of the limb, allowing the residual muscle power to move the arm. There is a motorized version, powered by the batteries of the wheelchair, which is actuated by microswitches on the forearm supports.