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XII. Muscular Dystrophy

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A Study of the Mechanism of Spinal Collapse in Duchenne Muscular Dystrophy

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Sponsor: *Regency Park Centre for Young Disabled*

Progress—A simple model consisting of a slender beam and support wires was constructed as an initial first model of the spine and its muscles. This model could give an indication of the sets of applied forces and/or moments that will restore as much as possible the spinal configuration. The model is solved by forming the stiffness matrix of the structure, K, and solving the load-displacement equation $P=Kd$. The difficulty is in accurately deducing the appropriate input data that would resemble the properties of the spine.

Data from literature give the stiffness matrix for some intervertebral joints (minus muscles). Therefore, it should be possible to form an approximate stiffness matrix of the whole column (minus muscles) and then form a modified stiffness matrix by including muscles modelled as springs or wires. Compare the results with the slender beam and support wires model. There is, however, a wide variation in stiffness matrix data from the literature. We are also modelling the spine by an anisotropic beam supporting a rigid body. We will look at the effect of a tilted pelvis and varying rigid body positions in a muscle-relaxed position, considering first only lateral bending, and deduce the equilibrium positions of the spine. Generally, we could expect three equilibrium positions. The model will be generalized and refined to include kyphotic curvature and sagital bending, external and internal loads, and constraints of movements.

Forearm Levitation

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Purpose—A mechanism has been designed by the Volunteers for Medical Engineering Inc. (VME) which supports the forearm of a person who does not have enough strength to raise it without some help. The device essentially levitates the arm, closely counter-balancing its weight so that only minimal force is required to raise or lower the arm.

The original design was made for a young man with muscular dystrophy and he has agreed to evaluate it for the VME.

Progress—The overall layout of the mechanism has been completed and the detail drawings for the fabrication of the prototype are under way. The fabrication was started in July 1986 and the first prototype was ready for testing in the fall of 1986.

Preliminary Results—A preliminary analysis of the mechanism is complete. A more detailed structural analysis will be started shortly and will be completed in time for iteration of the design before the unit fabrication is complete.

The detailed purchased parts list is complete and orders will be placed for two assemblies, enabling the VME volunteers to have a unit for continuing evaluation while the client

is using the second unit.

Future Plans—The unit now being fabricated is custom made for the client and will be useable only for others whose needs are similar and whose size and weight are similar. It is our goal to have the design computer database iteratable so that customization may be accomplished in a timely manner and the details rapidly updated for the new configuration. This would be readily accomplished because the present database is on the Interactive Graphs (IAG) computer system.

The Role of Spinal and Abdominal Muscles as Etiological Factors in Scoliosis in Neuromuscular Disorders

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Purpose—The purpose of our study is to determine whether computerized tomography of spinal and abdominal muscles in the dorsal and lumbar regions can be correlated with the degree of spinal curvatures in the neuromuscular disorders of Duchenne muscular dystrophy and spinal muscular atrophy, and whether this can be a prognostic indication of severity and speed of development of scoliosis.

Progress—There is a difference, in the pattern of muscle loss and/or replacement with either fat or fibrous tissue in both spinal and abdominal muscles, between boys who sit with a straight spine or a lordotic stance and develop only minimal scoliosis and those who are more kyphotic and develop severe scoliosis. This difference is detectable on C.T. scans.

The course of Duchenne muscular dystrophy is one of relentless progression of all voluntary muscles. While the subjects remain ambulant, there is an increase in the lumbar lordosis which is related to the weakness of the pelvic girdle musculature and a shift in center of gravity. Between the ages of 8 and 12, the children may cease to ambulate and become wheelchair bound. From this time, the greater proportion of them will develop a scoliosis or a kyphoscoliosis. In the case of spinal muscular atrophy, scoliosis is usual and may occur earlier or later according to the severity of the child's condition.

Preliminary Results—While the etiology of scoliosis in neuromuscular disorders has not been elucidated, it has generally been attributed to asymmetrical weakness of the muscles which support the spine. Wilkins and Gibson ("The patterns of spinal deformity in Duchenne muscular dystrophy." *J Bone and Joint Surg* 58-A, 24-32, 1976) in the clinical and roentgenographic study of 62 patients with Duchenne dystrophy observed that some patients with hyperextended spines had comparatively little scoliosis, while those who sat with a kyphotic posture tended to develop severe scoliosis. At the Regency Park Centre we have confirmed these findings clinically over a number of years.

Further work by Koreska et al. ("Biomechanics of the lumbar spine and its clinical significance." *Orthopaedic Clinics of North America* 8, 121-133, 1977) on the biomechanics of the lumbar spine and its clinical significance suggested that locking the posterior facets by trying to induce a "built in" lordotic posture in a wheelchair would reduce or prevent scoliosis. This latter hypothesis has not stood the test of time, however, and further work by Seeger & Sutherland ("Lumbar extension in Duchenne muscular dystrophy: Effect on lateral curvature." *Arch Phys Med and Rehabil*, 66, 236-238, 1985) did not support that finding. Earlier work by Stern et al. ("Progression of muscular dystrophy assessed by computed tomography." *Developmental Medicine and Child Neurology*, 26, 569-573, 1984) has shown that computer tomography is an accurate means of detecting the changes in dystrophic muscle and over a longi-

tudinal period of time can accurately reflect the changes in the muscle.

Future Plans—We propose to include 20 subjects with Duchenne dystrophy or spinal muscular atrophy in the study. Two scans will be taken, one through the dorsal region and one through the lumbar region. This gives good access to spinal and abdominal muscles. Density readings would be taken on both sides and compared. The density readings are a good indication of loss of muscle fibres. In each case the readings will be correlated with the degree of spinal X-rays which have already been taken to monitor the scoliosis. Similar readings would be repeated in 12 months to monitor the changes in the muscles.

It is hoped that these observations will produce some insight into the etiological factors of the scoliosis, and possibly suggest new strategies to approach the problem of prevention and treatment.

A Random Crossover Trial of Respiratory Muscle Endurance Training in Duchenne Muscular Dystrophy

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Purpose—The aim of the project is to test the hypothesis that regular ventilatory muscle training over sufficiently long periods will show statistically significant improvement in ventilatory muscle endurance in children with Duchenne muscular dystrophy. It is proposed to carry out the ventilatory muscle training by the use of computer games which will be adapted to be activated by the inspiratory efforts of the subjects.

Improvement of ventilatory muscle endurance could, in the longer term, lead to a slowing in the rate of pulmonary function deterioration, which occurs in Duchenne dystrophy, and delay the onset of respiratory failure.

Progress—The Electrical Engineering Department of the University of Adelaide has developed a means of monitoring the degree of breathing effort (or air velocity) during a training session. This unit uses a thermistor in a tube through which the person breathes. In order to keep the children's interest up, they will be able to play video games for as long as they breathe hard enough. If they do not satisfy this criterion, the joystick controlling the game will be temporarily disconnected. A bar of light-emitting diodes indicate how well the children are doing and how close they are to be disconnected.

The 15 boys involved in this study will be divided into two study groups with one group playing the video games and the other not. This situation will be reversed after 6 months. Each individual will have pulmonary function tests including measurement of inspiratory endurance at the beginning of the trial, at 6 months, and at 12 months.