XII. Respiration (Muscular Dystrophy)

Inspiratory Muscle Fatigue as a Cause of Respiratory Insufficiency in the Muscular Dystrophies and other Neuromuscular Diseases

Francois Haas, Ph.D.
New York University Medical Center
Department of Rehabilitation Medicine
New York, New York 10016

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Muscular Dystrophy Association

The inability to meet oxygen demands during work levels because of ventilatory muscle fatigue can result in a decrease in the efficiency of a rehabilitation program. Results from isotonic exercises to increase respiratory muscle endurance have been equivocal.

The use of isokinetic (resistive) exercise has not been fully explored as a technique for respiratory muscle readaptation. Consequently, we have devised an isokinetic exercise program for inspiratory muscles to be used in patients with neuromuscularly-induced weakness of respiratory muscles. Improvement will be assessed in terms of isometric pressure generation, peak airflow, and maximum breathing capacities.

Respiratory Load Compensating Mechanisms in Muscular Dystrophy

Kenneth Axen, Ph.D.
New York University Medical Center
Department of Rehabilitation Medicine
New York, New York 10016

Sponsor: National Institute of Handicapped Research
Muscular Dystrophy Association

Although it is well established that respiratory problems are a leading cause of disability and death in the muscular dystrophies (MD), the mechanisms underlying respiratory insufficiency remain unclear. In light of recent work identifying respiratory sensory and central abnormalities, we propose that these problems represent a combination of respiratory sensory, central, and motor dysfunctions.

It is proposed to test this hypothesis using the loaded breathing technique in which external mechanical loads are imposed noninvasively at the mouth to impede inspiration. Loads disrupt the usual relationship between the effort and corresponding movement of the respiratory system and, in doing so, activate those mechanisms that normally defend ventilation in the face of naturally occuring loads. This technique thus assesses the combined action of respiratory sensory, central, and motor functions during spontaneous breathing.

Despite these obvious advantages, the study of loaded breathing in MD has been precluded in the past by: (i) the lack of an adequate analytical method of identifying the action of load-compensating mechanisms; and (ii) the lack of an adequate normal standard. As is described in the literature, we have overcome both these problems by deriving a mathematical model to identify load-compensating mechanisms, and by applying it to interpret data obtained from large groups of normal and quadriplegic people. This information will be used to: (i) investigate the relationship between load-compensation and severity of disease; (ii) evaluate residual load compensation; and (iii) quantify the deficit in load compensation in groups of subjects with MD. We believe these new findings will be of fundamental importance in the understanding of ventilatory regulation in the muscular dystrophies and, ultimately, in the design of appropriate rehabilitative regimens to alleviate the tragic respiratory problems associated with these diseases.