

Poster Abstract: Clinical

Motor Function and Respiratory Capacity in Patients with Late-Onset Pompe Disease

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BACKGROUND

The relationship between skeletal muscle strength and respiratory dysfunction in Pompe disease has not been examined by quantitative methods. We investigated correlations among lower extremity proximal muscle strength, respiratory function, and motor performance.

MATERIALS AND METHODS

Concentric strength of the knee extensor and flexor muscles was measured with a dynamometer, and pulmonary function was evaluated using spirometry in 7 patients with late-onset disease. The 6-minute walk test and the 4-step stair-climb test were used for assessing aerobic endurance and anaerobic power, respectively.

RESULTS

Anaerobic motor performance correlated with strength of both thigh muscles. Respiratory function did not correlate with either muscle strength or motor function performance.

CONCLUSIONS

Respiratory and lower extremity proximal muscles could be differentially affected by the disease in individual patients. Motor performance is influenced by thigh muscle strength and is less dependent of respiratory capacity in our cohort of ambulatory patients.

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