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Authors

Kimonis, Virginia
Shah, Jeet
Bhatnagar, Caleb
[et al.](#)

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192**Safety and effectiveness of resistance exercise training in a pilot study of patients with late onset Pompe disease**

Virginia Kimonis, Jeet Shah, Caleb Bhatnagar, Bhumi Ramani, Vince Caiozzo, *UC Irvine, Orange, CA, United States*

Pompe Disease is a rare inherited disorder associated with muscle weakness and respiratory insufficiency which can affect all ages, ethnicities, and gender. It is caused by the accumulation of glycogen primarily in cardiac and skeletal muscle. There are two forms of the disease, classic infantile form and juvenile/adult form. The purpose of this study is to evaluate the effectiveness of the supervised resistance training program on muscle strength, functional capacity and body composition in patients with late onset Pompe disease. The study includes 10 patients over a 32 week study of increased resistance exercise 3 times a week in a gym with a personal trainer. The patients also undertook respiratory muscle exercise training daily with increasing resistance every 2 months. Each patient served as their own control (Weeks 1 - 8 baseline period) and subsequently physical strength, stamina and respiratory function was tested with the Biodex and hand held dynamometers, 6 minute walk test (6MWT), maximum/minimum inspiratory pressure (MIP/MEP), SF-36 questionnaire, muscle volume and texture change using MRI, and blood and urine collections of tetrasaccharide. Throughout the study, we found that there was an overall improvement in the patient's muscle strength by Biodex dynamometry, MRC scores and 6MWT. For example, knee and elbow extension torque improved by a mean of 21.6% and a 41.6% respectively. The 6MWT showed an average improvement of 6.58%, and total MRC scores showed an improvement of 2.1% from initial to final visit. There was a marked improvement in the respiratory parameters in the majority of patients the MIP showed an increase of 25% while MEP increased by 10%. Overall, patients also had an improved sense of well being with very few adverse side effects.

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