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## Moderate-intensity aerobic exercise improves physical fitness in bethlem myopathy.

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### **Abstract**

**INTRODUCTION:** Bethlem myopathy is caused by dysfunctional collagen VI assembly, leading to varying degrees of hyperlaxity, contractures and muscle weakness. Previous studies demonstrate that cardiovascular training is safe and beneficial in patients with myopathies. However, exercise exacerbates the dystrophic phenotype in collagen VI-knockout mice.

**METHODS:** Six men with Bethlem myopathy were included (4 training; 2 controls). After training, 2 patients detrained. Patients performed 10 weeks of home-based, moderate-intensity exercise monitored by a pulse-watch. The primary outcome was change in peak oxygen uptake ( $VO_{2peak}$ ). Secondary outcomes were performances in functional tests.

**RESULTS:**  $VO_{2peak}$  improved in the training group (16%,  $P = 0.017$ ). Detraining led to regression of  $VO_{2peak}$  toward baseline values (-8%;  $P = 0.03$ ). No change was seen in the control group (-7%;  $P = 0.47$ ). Performance in functional tests did not change significantly. Creatine kinase values were stable during the study.

**CONCLUSIONS:** Moderate-intensity exercise seems to safely improve oxidative function in patients with Bethlem myopathy. *Muscle Nerve* 60: 183-188, 2019.

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**KEYWORDS:** Bethlem myopathy; collagen VI; moderate-intensity exercise; muscular dystrophy; myopathy; peak oxygen uptake; physical exercise; training intervention

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