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Format: Abstract


In vivo characterization of skeletal muscle function in nebulin-deficient mice.

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Abstract

INTRODUCTION: The conditional nebulin knockout mouse is a new model mimicking nemaline myopathy, a rare disease characterized by muscle weakness and rods within muscle fibers. We investigated the impact of nebulin (NEB) deficiency on muscle function in vivo.

METHODS: Conditional nebulin knockout mice and control littermates were studied at 10 to 12 months. Muscle function (force and fatigue) and anatomy (muscles volume and fat content) were measured in vivo. Myosin heavy chain (MHC) composition and nebulin (NEB) protein expression were assessed by protein electrophoresis.

RESULTS: Conditional nebulin knockout mice displayed a lower NEB level (-90%) leading to a 40% and 45% reduction in specific maximal force production and muscles volume, respectively. Nebulin deficiency was also associated with higher resistance to fatigue and increased MHC I content.

DISCUSSION: Adult nebulin-deficient mice displayed severe muscle atrophy and weakness in vivo related to a low NEB content but an improved fatigue resistance due to a slower contractile phenotype.

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KEYWORDS: in vivo; fatigue; muscle wasting; muscle weakness; nemaline myopathy; neuromuscular disorder

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