Opa1 overexpression ameliorates the clinical phenotype of two mitochondrial disease mouse models

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Mitochondrial diseases are highly heterogeneous disorders characterized by faulty oxidative phosphorylation. OPA1 is a protein of the inner mitochondrial membrane, which shapes mitochondrial cristae, the structures where the respiratory complexes are located, thus regulating the efficiency of the respiratory chain.

Civiletto et al. have exploited this concept to investigate whether moderate Opa1 overexpression in tissues could ameliorate the clinical and biochemical phenotype of two mouse models of severe mitochondrial disease, characterized by deficiency in respiratory complex I and IV, respectively. These biochemical defects lead to reduced motor and coordination skill, reduced lifespan and reduced activity of the respective respiratory complexes.

The mice with increased OPA1 levels showed a marked improvement in both clinical and biochemical defects of the two mouse models. These effects were related to the correction of the mitochondrial ultrastructure and stabilization of the respiratory complexes and supercomplexes. By identifying OPA1 as a new potential target, this study opens new avenues towards evidence-based effective therapy for mitochondrial disorders.

More information: "Opa1 Overexpression Ameliorates the Phenotype of Two Mitochondrial Disease Mouse Models," *Cell Metabolism*, Volume 21, Issue 6, 2 June 2015, Pages 845-854, ISSN 1550-4131,

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