

Physiological impact and disease reversion for the severe form of centronuclear myopathy linked to Dynamin

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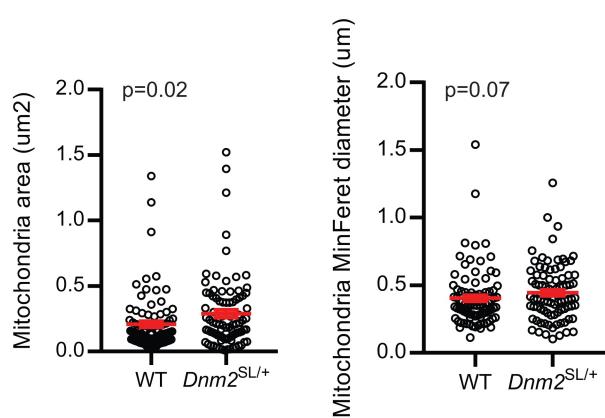
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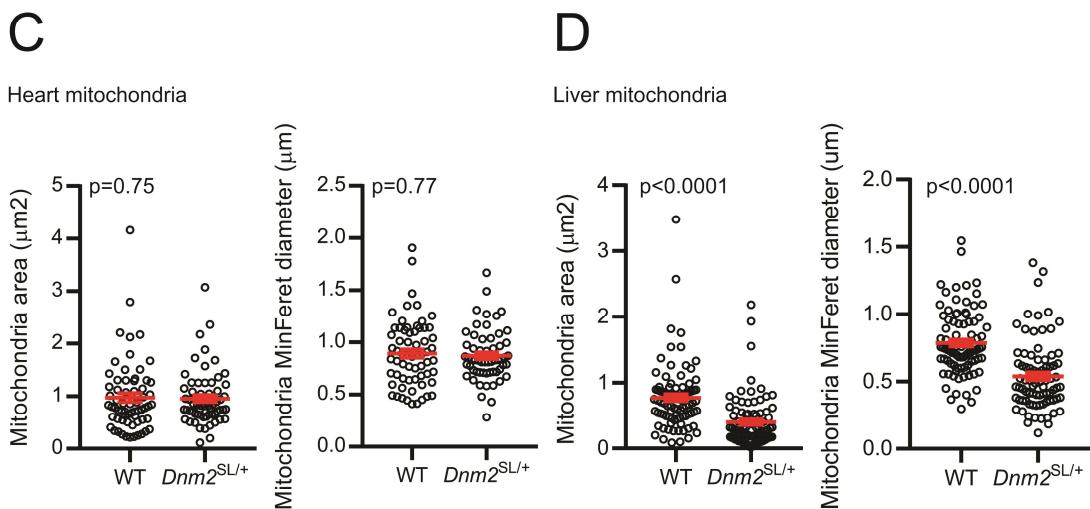
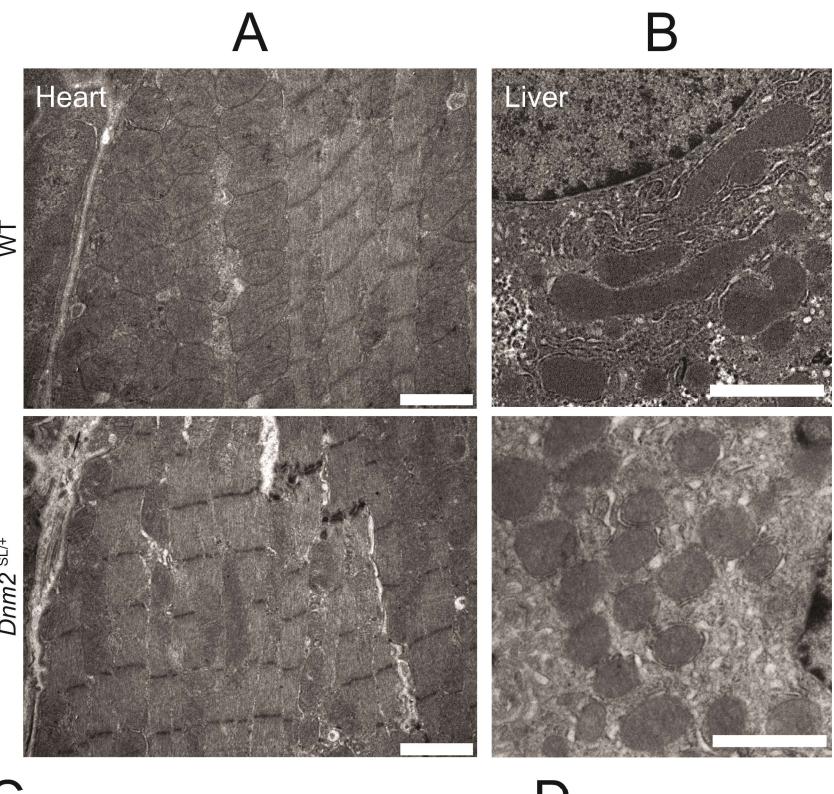
equal contributors

A

Skeletal muscle mitochondria

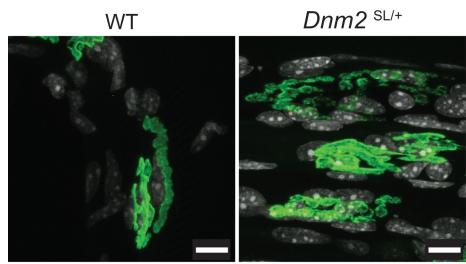


Supplementary Figure 1. Increased area of $Dnm2^{SL/+}$ tibialis anterior mitochondria.

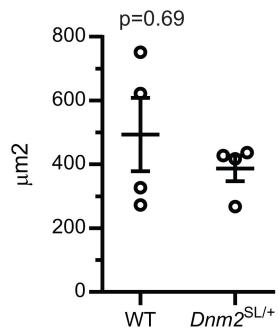


Supplementary Figure 2. Analysis of heart and liver mitochondria. (A-B) Representative images from heart (A) and liver (B) mitochondria (Scalebar = 2 μm). (C-D) Quantification of area and Minimum Feret diameter in heart (C) and liver (D) reporting differences in liver mitochondrial size. (In both cases 60 $\leq n \leq$ 88 mitochondria (2 animals, 3 different regions of interest), Mann Whitney test).

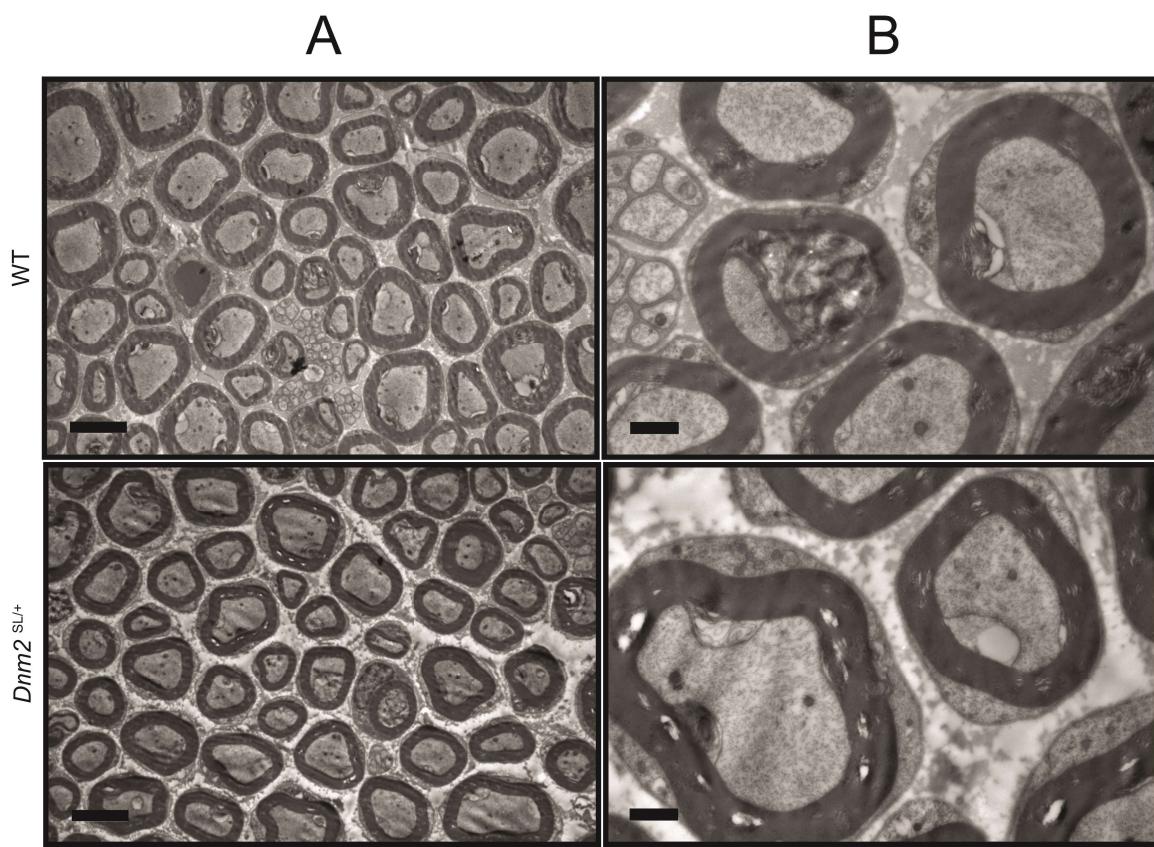
A



B



Supplementary Figure 3. Neuromuscular junction staining. (A) Representative images of α -bungarotoxin staining (in green) and DAPI (in gray) in fibers from 8w old WT and $Dnm2^{SL/+}$ mice. Scale bar=10 μm . (B) Quantification of neuromuscular junction area (n=4).



Supplementary Figure 4. Transmission electron microscopy images from sciatic nerve. (A) Transversal sections of sciatic nerve from 8w old WT and *Dnm2*^{SL/+} mice. Scale bar=5 μ m. (B) Higher magnification of the same sciatic nerves from 8w old WT and *Dnm2*^{SL/+} mice. Scale bar=1 μ m.

Figure 1F

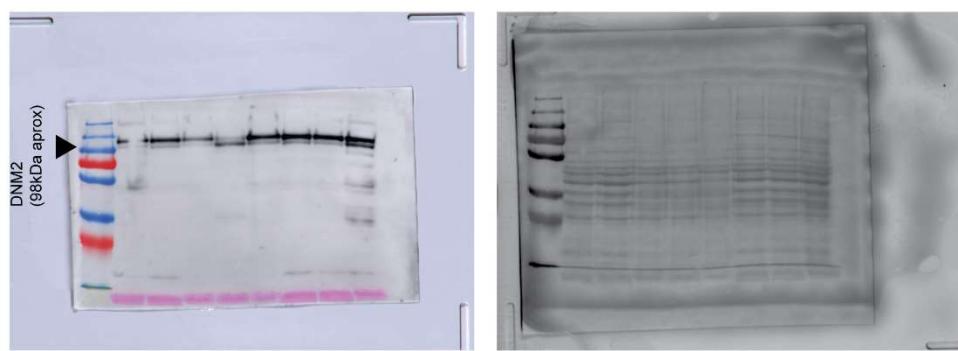
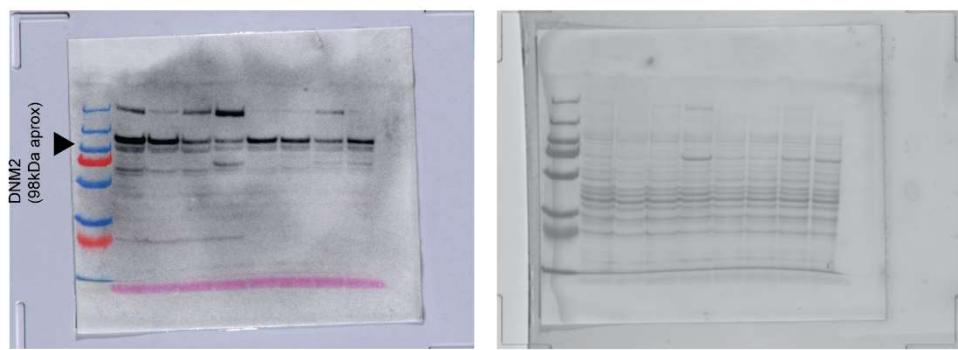


Figure 5H



Supplementary figure 5. Uncropped full-length Western blot images.

Supplementary movie 1. Hanging test performance of *Dnm2*^{SL/+} treated with ASO. Video showing two 8 weeks old *Dnm2*^{SL/+} mice untreated and treated with ASO performing hanging test.