Supplementary Figures


Suppl Fig. 1. Cell culture and plantaris muscle characteristics in Lmna ${ }^{\mathrm{H} 222 \mathrm{P}}$ mutation. (A) Confocal immunofluorescence images of myosin (MF20, green) in WT and LMNA ${ }^{\text {H222P }}$ cells after 3 days of differentiation. Nuclei are stained with Hoechst (blue). Scale bar=50 $\mu \mathrm{m}$. (B) Fusion index in WT and LMNA-CMD mutant cells after 3 days of differentiation. Values are expressed as means $\pm$ SEM. (C) Plantaris muscle mass normalized by body mass from WT and Lmna ${ }^{\text {H222P }}$ mice in control and after 4 -weeks FO. * $\mathrm{p}<0.05$ versus control conditions. Values are expressed as means $\pm$ SEM.

A


Suppl Fig. 2. Nuclear deformations in Lmna+/DK32 mice following functional overload. (A) Confocal immunofluorescence images of nuclei (Hoechst, white) in WT and Lmna+/AK32 in control and after 7-days FO. (B) Nucleus deformations in WT and Lmna+/DK32 in control and after 7 -days FO. Values are means $\pm$ SEM, $n \geq 230$ nuclei/condition.

A


B

C D


Suppl Fig. 3. Histological data from muscle biopsies of controls and a patient with EDMD.
(A) Immunofluorescence images of PAX7+ (red) and laminin (green) in muscle section from a control 4-day-old boy, a control 33-year-old man and an EDMD patient 59-year-old with heterozygous $L M N A^{H 222 P}$ mutation. Nuclei are stained with Hoechst (blue). Scale bar: $30 \mu \mathrm{~m}$. (B) Immunofluorescence images of YAP (red) and laminin (green) in muscle section from section from a control 4-day-old boy, a control 33 -year-old man and an EDMD patient 59 -year-old with heterozygous LMNA ${ }^{\text {H222P }}$ mutation. Nuclei are stained with Hoechst (blue). Scale bar: $50 \mu \mathrm{~m}$. (C, D) Quantification YAP+ cells per fibre and YAP+ cells per nucleus in controls and EDMD patient.

