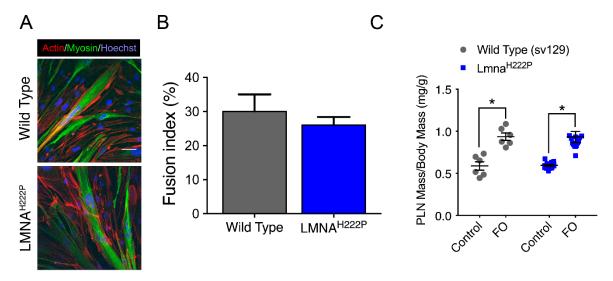
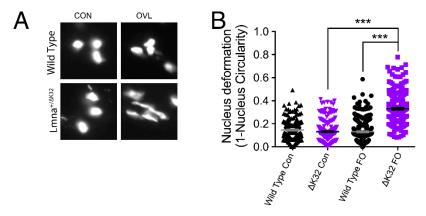
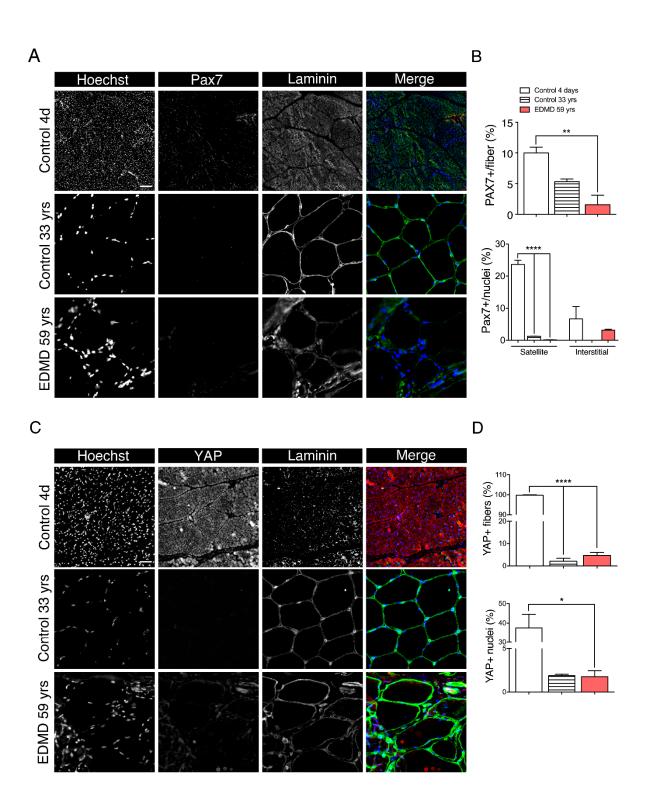
## **Supplementary Figures**



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



Suppl Fig. 2. Nuclear deformations in Lmna+/ $\Delta$ K32 mice following functional overload. (A) Confocal immunofluorescence images of nuclei (Hoechst, white) in WT and *Lmna*+/ $\Delta$ K32 in control and after 7-days FO. (B) Nucleus deformations in WT and Lmna+/ $\Delta$ K32 in control and after 7-days FO. Values are means ± SEM, n≥230 nuclei/condition.



## Suppl Fig. 3. Histological data from muscle biopsies of controls and a patient with EDMD.

(A) Immunofluorescence images of PAX7+ (red) and Iaminin (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 *LMNA*<sup>H222P</sup> mutation. Nuclei are stained with Hoechst (blue). Scale bar: 30 μm. (B) Immunofluorescence images of YAP (red) and Iaminin (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 boy, a control 33-year-old man and an EDMD patient 59-year-old with heterozygous *LMNA*<sup>H222P</sup> mutation. Nuclei are stained with Hoechst (blue). Scale bar: 50 μm. (C, D) Quantification YAP+ cells per fibre and YAP+ cells per nucleus in controls and EDMD patient.