R-DMDdel52 rat model of Duchenne muscular dystrophy for the identification of Comp as biomarker of fibrosis

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Supplementary information

Figures S1, S2 and S3.
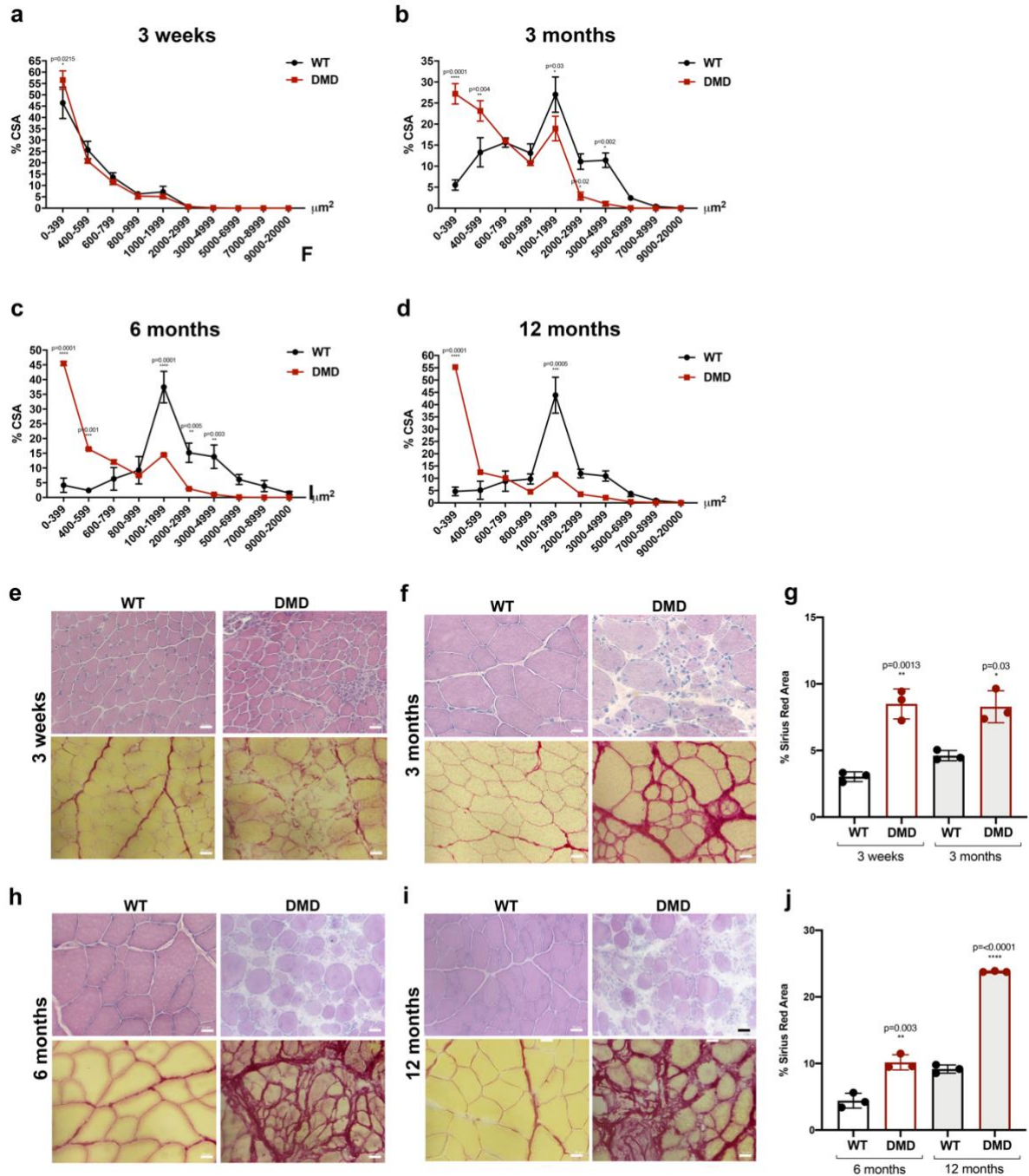
Figure S1

Fig.S1 Myofibre CSA of the diaphragm and morphological evaluation of TA muscle

a) to d) CSA of diaphragm of WT and R-DMDdel52 rats at 3 weeks (a), 3 months (b), 6 months (c) and 12 months of age (d). e) Hematoxylin and eosin (upper panel) and Sirius red (lower panel) staining of *tibialis anterior* of 3-week-old WT and R-DMDdel52 rats (scale bar = 50 μm). f) Hematoxylin and eosin (upper panel) and Sirius red (lower panel) staining of *tibialis anterior* of 3-month-old WT and R-DMDdel52 rats (scale bar = 50 μm). g) Quantification of fibrotic Area of E-F. g) Quantification of fibrotic Area of e-f. h) Hematoxylin and eosin (upper panel) and Sirius red (lower panel) staining of *tibialis anterior* of 6-month-old WT and R-DMDdel52 rats (scale bar = 50 μm). i) Hematoxylin and eosin (upper panel) and Sirius red (lower panel) staining of *tibialis anterior* of 12-month-old WT and R-DMDdel52 rats (scale bar = 50 μm). j) Quantification of fibrotic Area of h-i.
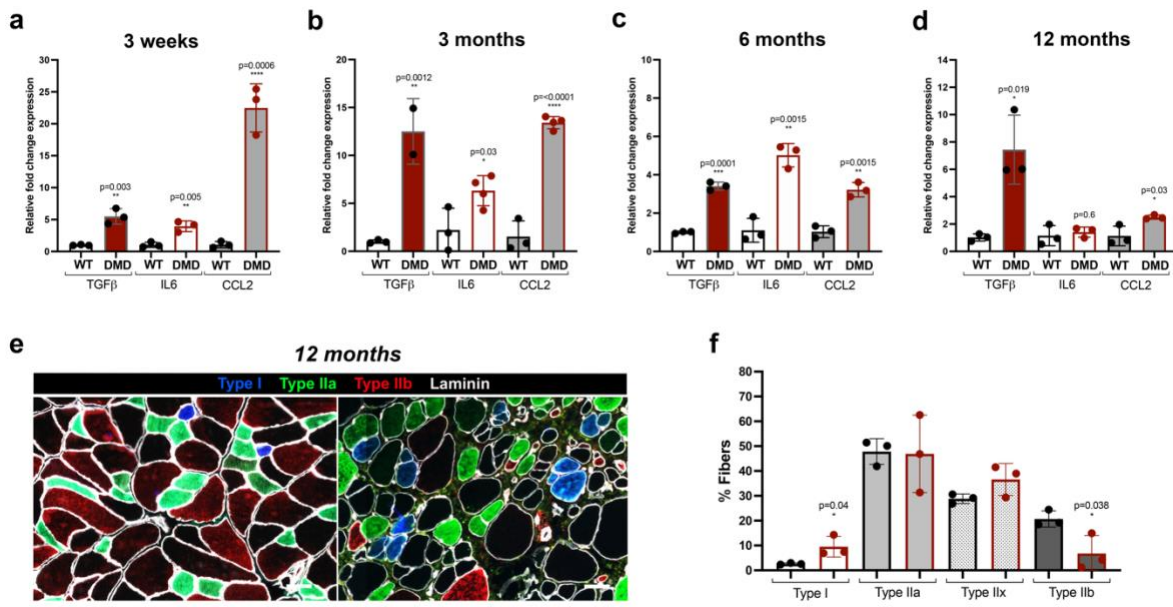
Fig. S2 | Inflammatory cytokine evaluation and fibre type switching in R-DMDdel52 rats.

a-d) qPCR analysis of inflammatory cytokines (*TGFβ, IL6,* and *CCL2*) on quadriceps of WT and R-DMDdel52 rats at 3 weeks (a), 3 months (b), 6 months (c) and 12 months (d). e) Immunofluorescence for Type I (blue), IIa (green), IIx (unstained, black), IIb fibres (red) in WT and R-DMDdel52 EDL at 12 months (scale bar = 50 μm). (f) Quantifications of the percentage of Type I, IIa, IIx, IIb fibres at 12 months of age in WT and R-DMDdel52 EDL muscles. Statistical analysis was performed with unpaired Student's t-test.
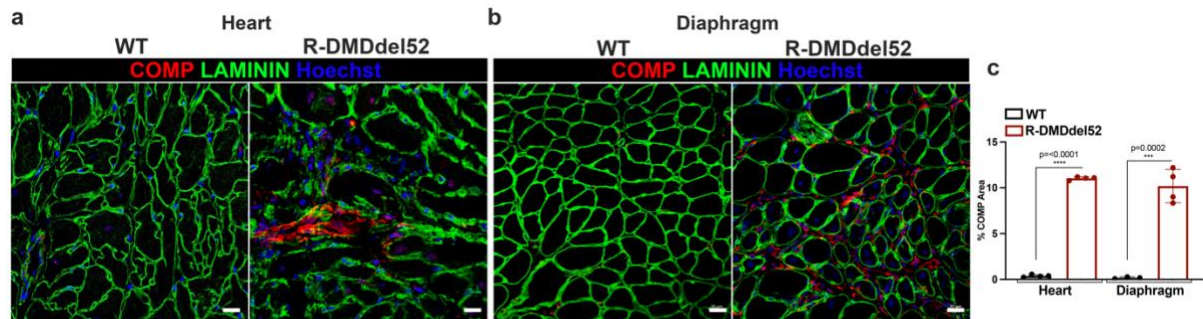
Fig. S3 | COMP expression in heart and diaphragm.

a) and b) Representative immunofluorescences of COMP expression in heart (a) and diaphragm (b) of WT and R-DMDdel52 rat at the age of 12 months. LAMININ is shown in green, and nuclei are stained with Hoechst (scale bar = 50 μm). c) Quantification of COMP positive area in heart and diaphragm at 12 months.