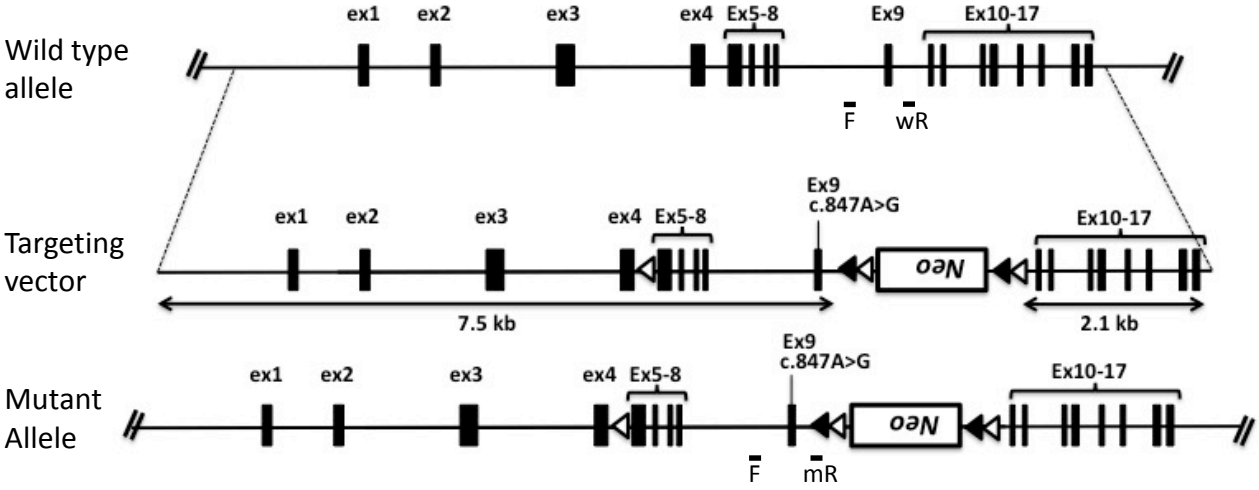
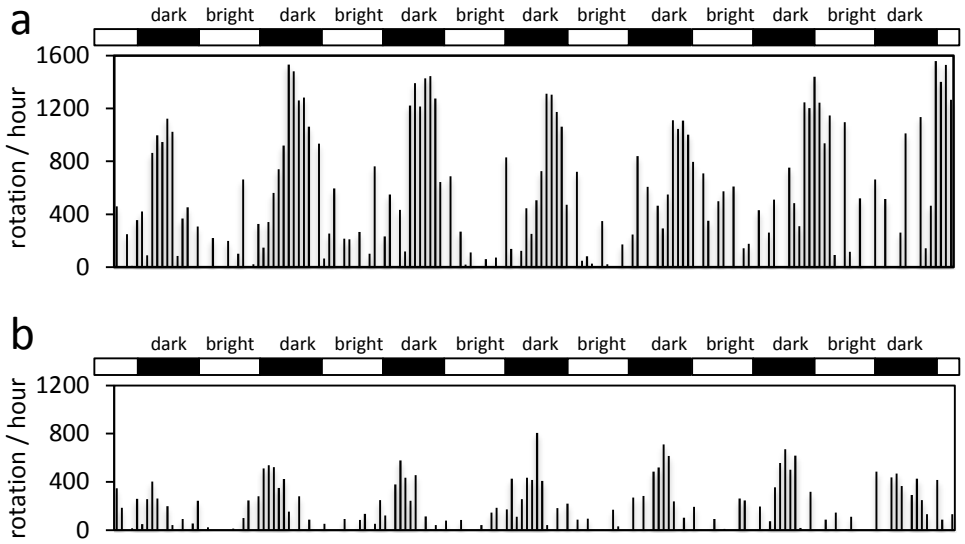
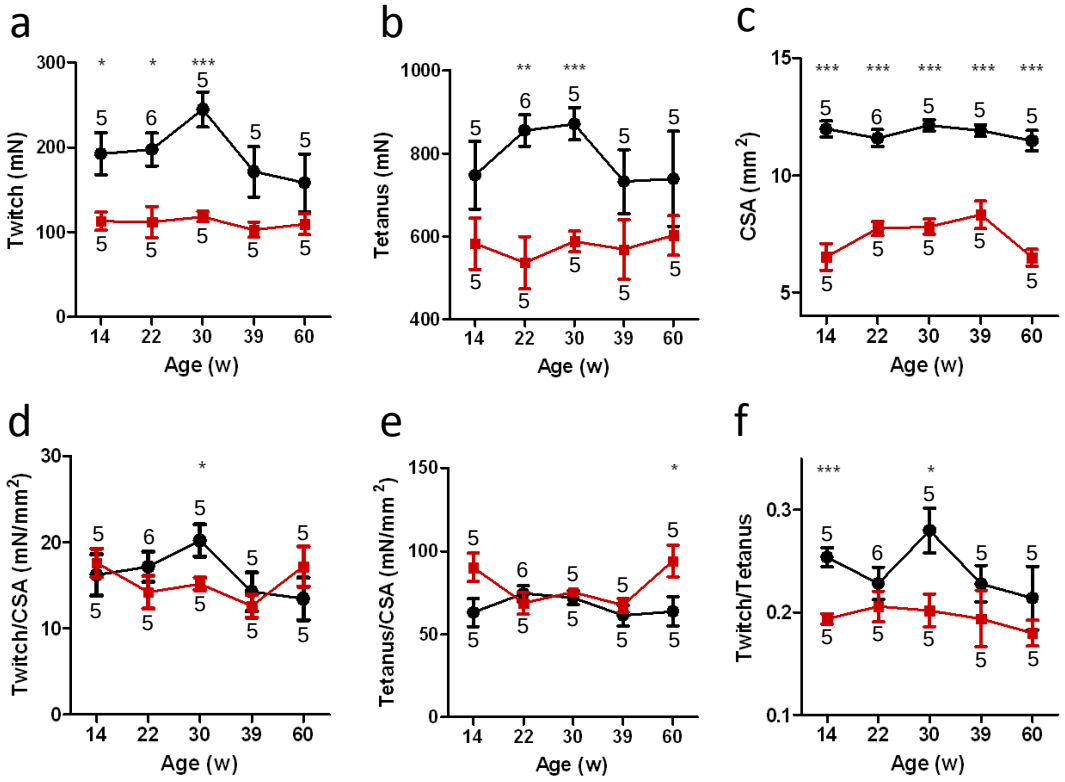


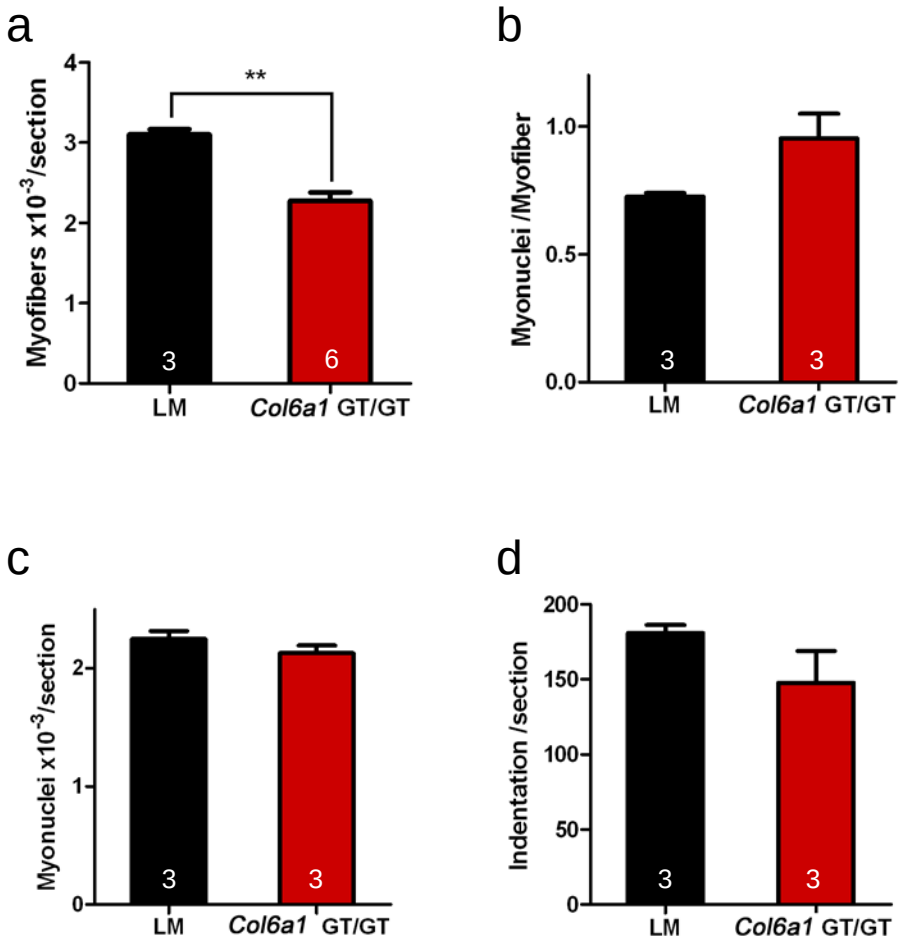
Supplementary Figure 1. Generation of the *Col6a1* mutant mice.
Schematic representation of the wild-type allele, the targeting vector, and the mutant allele of the mouse *Col6a1* gene. Primers for PCR genotyping (primer F, wR and mR) are shown as small closed boxes indicated by F, wR, and mR, respectively. Closed triangle, *FRT* site; open triangle, *loxP* site.



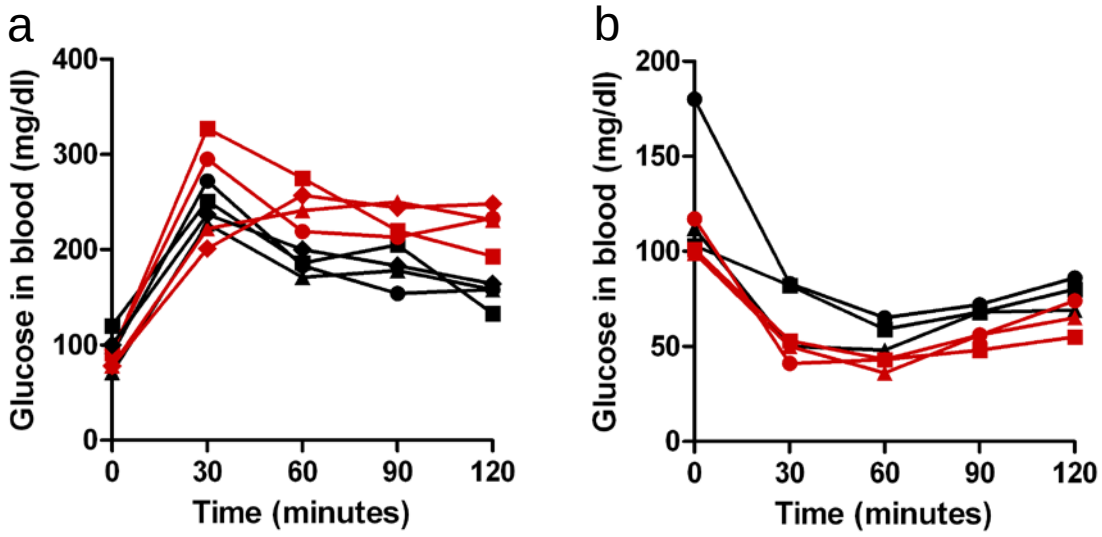
Supplementary Figure 2. Voluntary locomotion activities of the mice.
(a, b) Representative results of the voluntary locomotion of control littermate (LM) (a) and *Col6a1* GT/GT mice (b). Locomotion activities were measured for 7 days (dark, time in the dark; bright, time in the bright). Horizontal axis shows time with one unit of dark or bright bar is corresponding to 12 hours. Vertical axis shows rotations for 1 hour.



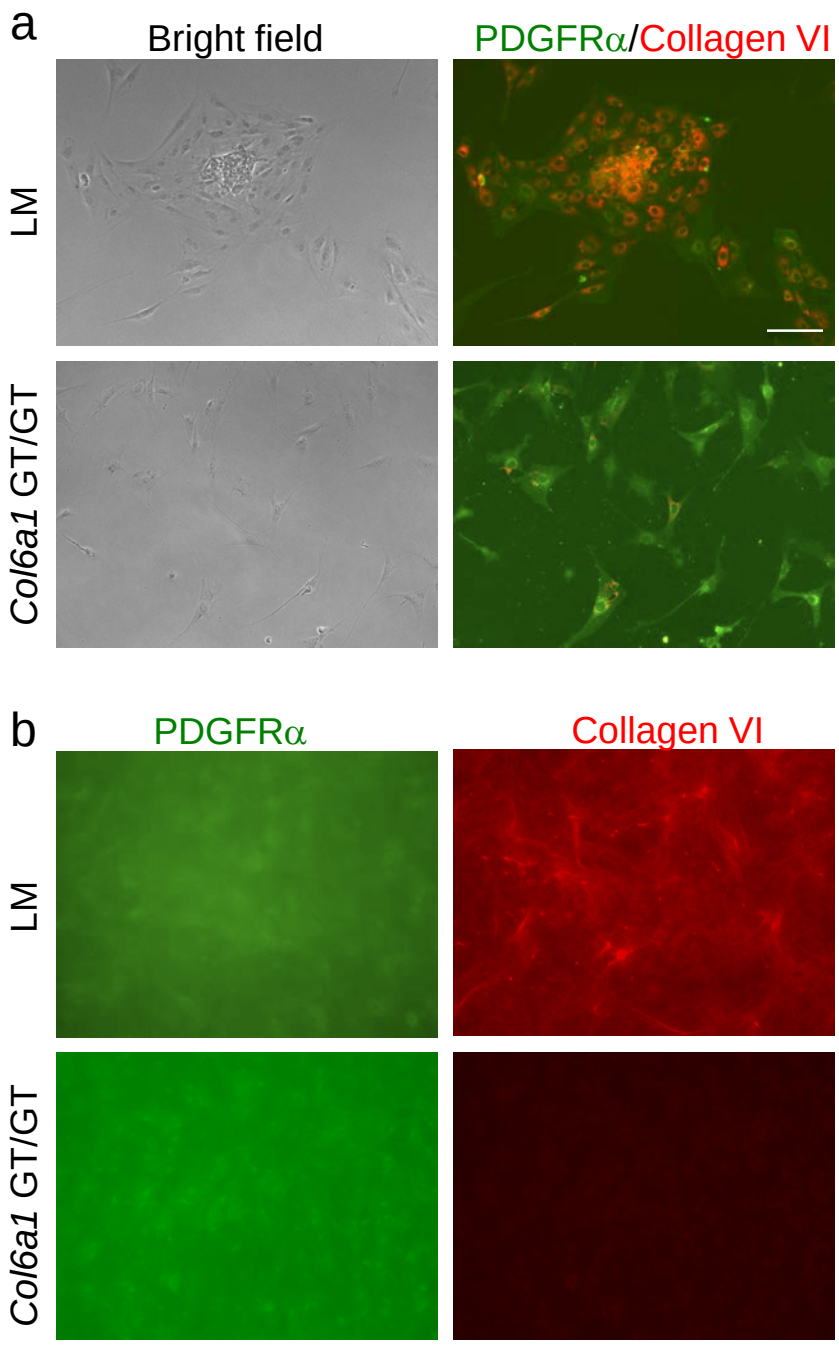
Supplementary Figure 3. Contractile properties of Gastrocnemius muscle, LM (black, n = 5-6 per aged group) and *Col6a1*GT/GT (red, n = 5 per aged group) (a) peak isometric twitch, (b) Maximal tetanic force, (c) Cross sectional area (CSA), (d) Specific isometric twitch (e) Specific tetanic force, (f) Twitch/tetanus ratio. Data represent the mean ± SEM. **p* < 0.05, **p < 0.01, ****p* < 0.001 on the paired *t*-test was used.



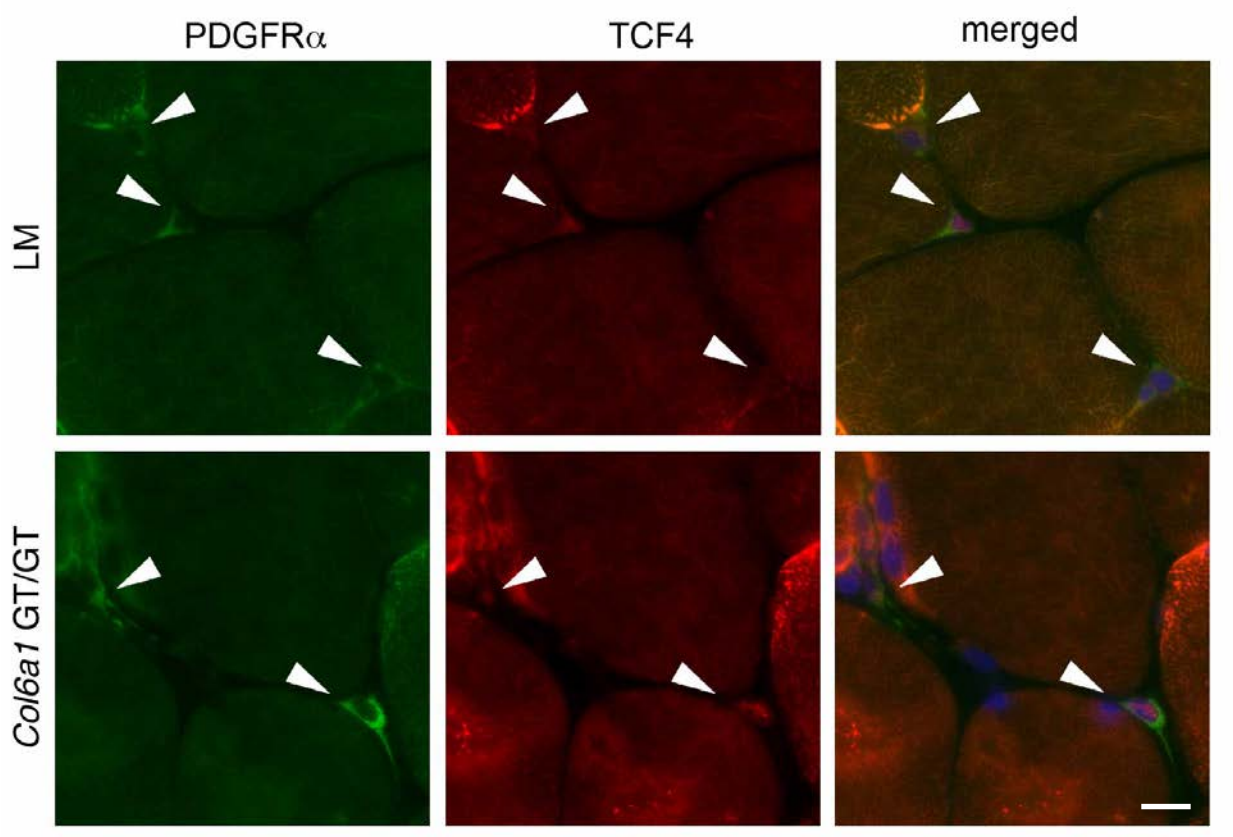
Supplementary Figure 4. Histological examination of TA muscles during neonatal stage. (a) Total myofiber number in TA muscles in control LM (black, n=3) and *Col6a1*$^{GT/GT}$ (red, n=3) at 21 days after birth. (b) The number of myonuclei per myofiber in control LM (black, n=3) and *Col6a1*$^{GT/GT}$ (red, n=3) at 21 days. (c) The number of myonuclei per section of TA muscles in control LM (black, n=3) and *Col6a1*$^{GT/GT}$ (red, n=3) at 21 days. (d) myofiber indentation in TA muscle sections in control LM (black, n=3) and *Col6a1*$^{GT/GT}$ (red, n=3) at 14 days.



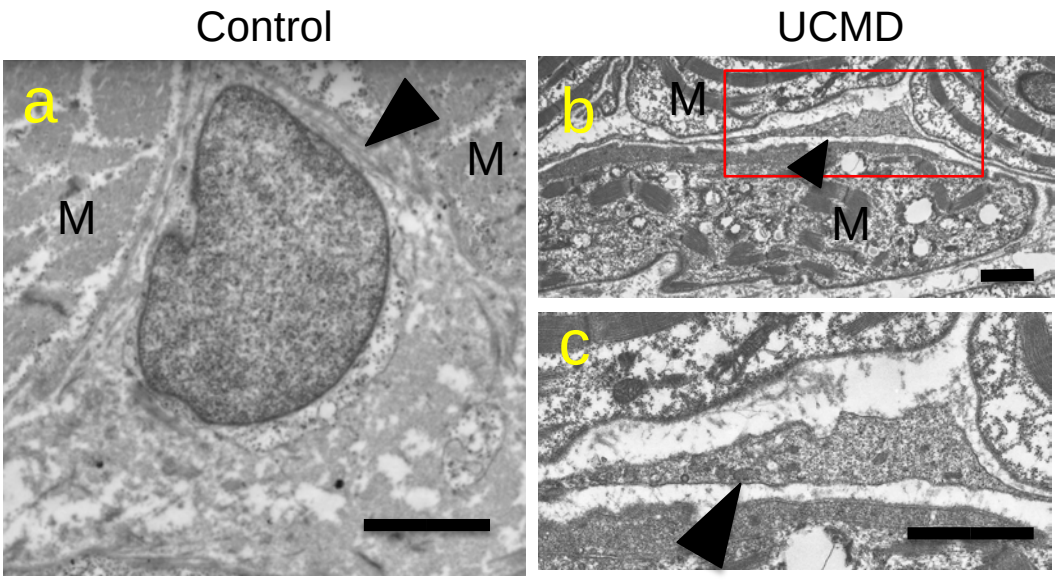
Supplementary Figure 5. Glucose tolerance and insulin resistance in LM (black) and *Col6a1* GT/GT mice (red). Mice were starved for 16 hours. (a) Blood glucose levels after glucose inoculation (40 mg) at time 0. (b) Blood glucose levels after injection of human insulin (28 mU) at time 0. The traces in three mice in each group are shown.



Supplementary Figure 6. PDGFRα-positive MPCs are source of collagen VI. (a) Isolated MPCs were cultured without ascorbic acid. In this condition, collagen VI was retained in the cytosol in MPCs from LM mice. (b)) MPCs were cultured with ascorbic acid. Collagen VI was secreted into extracellularly from MPCs derived from LM. Green, PDGFRα staining; red, collagen VI staining. Bar denotes 100 μm.



Supplementary Figure 7. Identification of MPCs as being dual positive to PDGFRα and Tcf4. Skeletal muscles from control and *Col6a1*$^{GT/GT}$ mice at 14 weeks were stained with PDGFRα (green), Tcf4 (red) and DAPI (blue). Arrowheads indicate MPCs. Bar denotes 10 μm.



Supplementary Figure 8. Morphology of interstitial cells is altered in UCMD muscle. (a) Endomysial non-muscle cell in skeletal muscle of control individual. It was round-shape with small cytoplasmic area and located close to myofibers (M). (b) Endomysial non-muscle cell in UCMD muscle. It has elongated. (c) Magnified image of an inset (red) in (b). There is space between MPC and myofibers. Bars denote 2 μm.