

**Supplementary information, Figure S4** 

**Fig. S4** DM1-associated phenotypes in adult DSM-TKO SC mice. **a** Genotyping analysis of TKO SC mice. **b** Immunofluorescent staining of dystrophin (green) in DSM-TKO and WT SC mice. Scale bars, 50  $\mu$ m. **c** ATPase staining showing the increased ratio and reduced CSA of type I myofiber (darker cells) in TKO SC mice (*n* = 4 per group). Unpaired Student's *t*-test, \*\**P* <

0.01. NS, no significant differences. Scale bars, 100 µm. **d** H&E staining of diaphragm showing increased fat cell invasion in TKO SC mice. Scale bars, 100 µm. **e** Histological analysis of small intestine. Boxes in left panels are magnified in right panels, showing disorganization of intestine villi in TKO SC mice. Scale bars, 100 µm. **f** IVPW-d thickness (diastole) of DSM-TKO and WT SC mice at different ages (n = 5 per group). IVPW: left ventricular posterior wall. Unpaired Student's *t*-test. NS, no significant differences. **g** Reduced ejection fraction in 12-month-old DSM-TKO SC mice (n = 5 per group). Unpaired Student's *t*-test, \*\*P < 0.01. NS, no significant differences. **h** Myocardial disarray shown in the 12-month-old TKO SC mouse. Scale bars, 50 µm.