

Fig. S1. Confocal images of YFP⁺-neurons from the CA2 region of the hippocampus (A) and midbrain (B) of sagittal brain cryosections of -/- mice with or without tamoxifen (Tx) administration and immunostained for Ranbp2. Sections were counterstained with DAPI. Immunostaining of Ranbp2 is lost from the nuclear rim of YFP⁺-neurons in Tx-treated mice and without causing overt morphological changes of YFP⁺-neurons. Legend: -/-, *SLICK-H::Ranbp2^{flox/flox}*; d10, day 10 post-tamoxifen administration; scale bars=50 μ m.

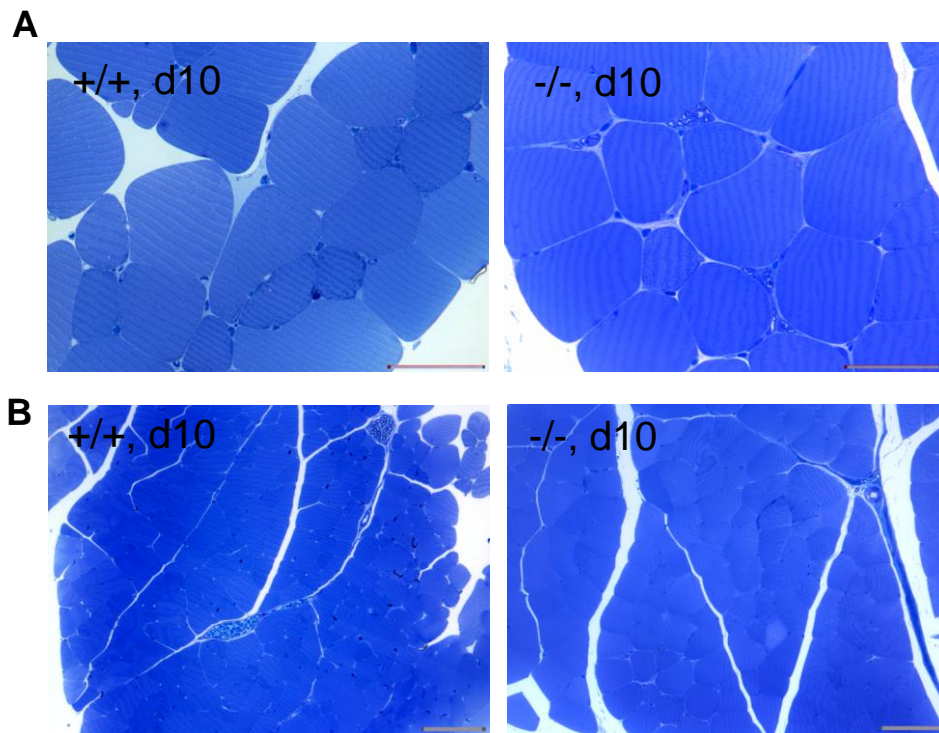


Fig. S2. Light microscopy of high (A) and low (B) magnification images of semithin sections of the gastrocnemius muscle stained with crystal violet of +/+ and -/- mice. No signs of denervation atrophy and muscle wasting is observed between genotypes. Legend: +/+, *SLICK-H::Ranbp2*^{+/+}; -/-, *SLICK-H::Ranbp2*^{flox/flox}; d10, day 10 post-tamoxifen administration; scale bars=50 μm.

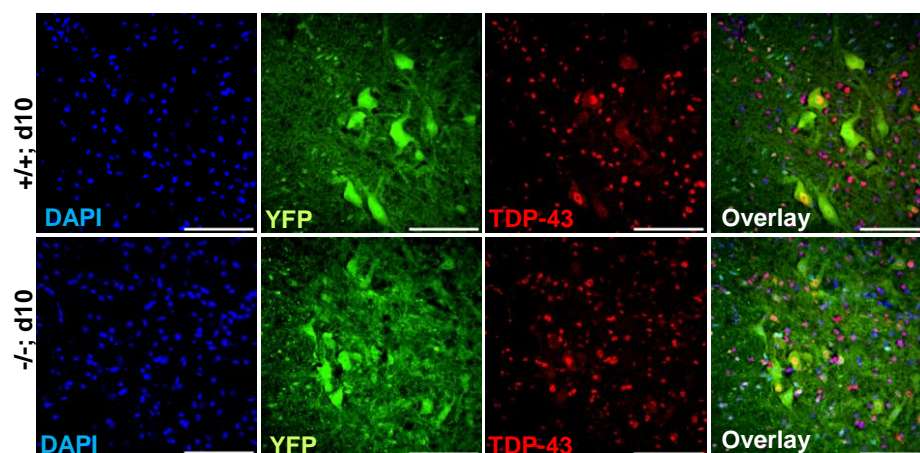


Fig. S3. Confocal images of YFP⁺-motoneurons of the anterior horn of the lumbar spinal cord immunostained for TDP-43. Sections were counterstained with DAPI. TDP-43 is localized to the nucleus of YFP⁺-motoneurons of +/+ and -/- mice. Legend: +/+, *SLICK-H::Ranbp2*^{+/+}; -/-, *SLICK-H::Ranbp2*^{flox/flox}; d10, day 10 post-tamoxifen administration; scale bars= 100 μ m.

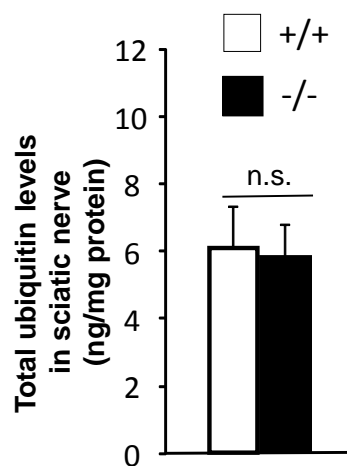


Fig. S4. Total ubiquitin levels are similar in the sciatic nerve of +/+ and -/- mice at d10. Data are expressed as mean \pm sd; $n=4$ mice/genotype; Student's t -tests; Legend: +/+, *SLICK-H::Ranbp2*^{+/+}; -/-, *SLICK-H::Ranbp2*^{fllox/fllox}; d10, day 10 post-tamoxifen administration; n.s., non-significant.



Movie S1. Locomotor behavior of *SLICK-H::Ranbp2^{flx/flx}* (-/-) mice at day 9 post-tamoxifen administration.