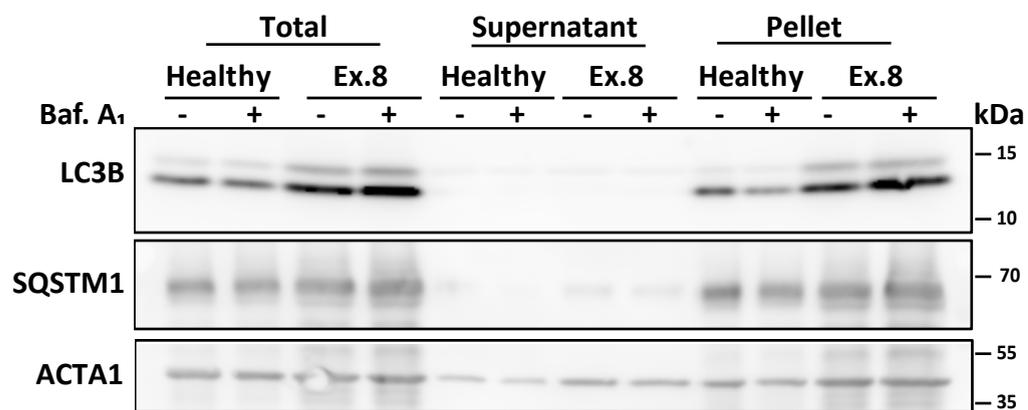
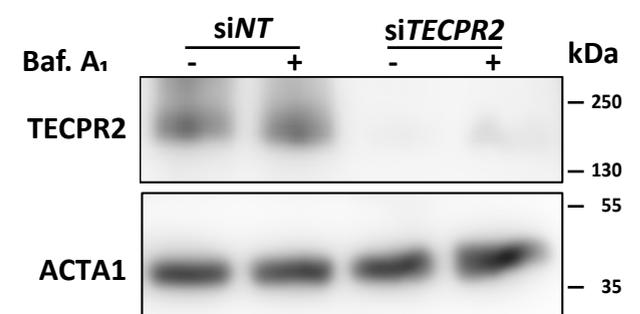


**A****B**

**Figure S1** - Accumulation of autophagy markers in primary fibroblast cells under basal conditions. **(A)** Primary fibroblast cells derived from healthy individual and from SPG49 patient with mutation on exon 8 were grown in complete medium till confluency and treated for last 4 h with 0.1 μM bafilomycin A<sub>1</sub> (Baf. A<sub>1</sub>) where indicated. Total protein extracts were separated by ultracentrifugation into supernatant and pellet fractions, and then analyzed by western blotting for TECPR2, LC3B and SQSTM1. **(B)** BJ primary fibroblast cells were transfected with siNT or siTECPR2 using Darmophact transfection reagent for 72 h with 0.1 μM bafilomycin A<sub>1</sub> for four last hours where indicated. Then cells lysate was subjected to the western blot analysis and the efficiency of siTECPR2 was detected by anti-TECPR2 antibody.