

6^{neo} Mouse Model

6^{neo} mice

The mouse model has a knockout in the acid α -glucosidase (GAA) gene, resulting in reduced GAA enzyme levels and thus a progressive accumulation of glycogen in different tissues.

Starting at 3 weeks:

- Reduced GAA activity in brain, spinal cord, liver, and muscle
- Glycogen accumulations in brain, spinal cord, heart, skeletal muscle, and diaphragm (PAS reaction)
- Reduced number of myofibrils
- Damaged muscle structure
- Reduced mobility

Later phenotypes:

- Reduced body weight
- Astrocytosis

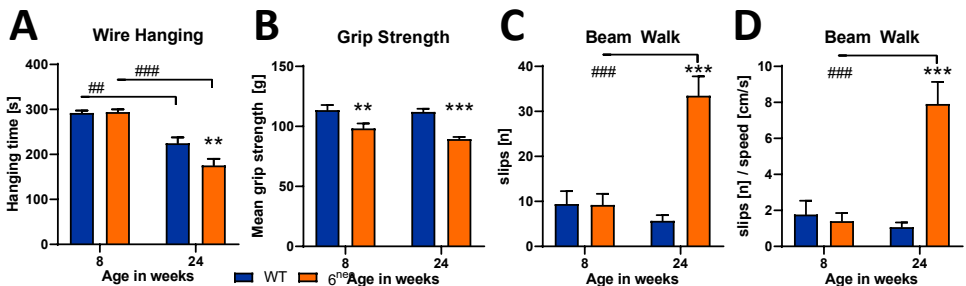


Figure: Muscle and motor deficits in the 6^{neo} mouse model of Pompe disease. 6^{neo} mice at the age of 8 and 24 weeks were tested for hanging time in the wire hanging test (A), for mean grip strength in the grip strength test (B) and for number of slips (C) and slips per speed (D) in the beam walk test (C+D). WT: n = 24; 6^{neo}: n = 48; mean + SEM; two-way ANOVA followed by Bonferroni's multiple comparison *post hoc* test; **/###p<0.01, ***/###p<0.001; *significance between genotypes; #significance between age groups.

References:

Raben et al., J Biol Chem. 1998 Jul 24;273(30):19086-92. doi: 10.1074/jbc.273.30.19086.

Lee et al., Mol Neurobiol. 2018 Jun;55(6):5299-5309. doi: 10.1007/s12035-017-0763-4.