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In vivo Animal Models

Pompe Disease

6^{neo} Mouse Model - 6^{neo} Mice

The mouse model has a knockout in the acida-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

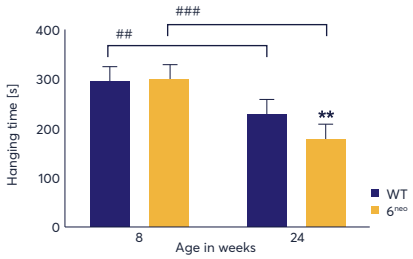
Later phenotypes:

- Reduced body weight
- Astrocytosis

Figure 1:
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.

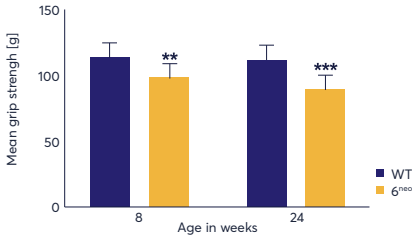
Wire Hanging

Figure 1: A



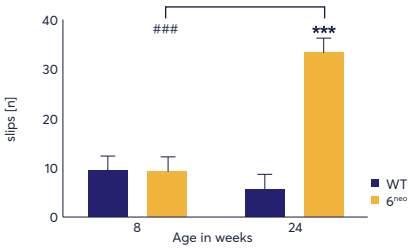
Grip Strength

Figure 1: B



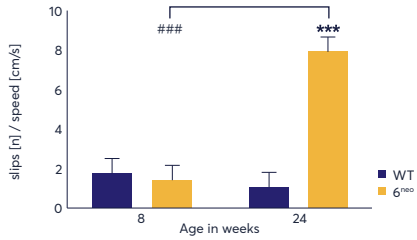
Beam Walk

Figure 1: C



Beam Walk

Figure 1: D



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.

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Discovery

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