



## 6<sup>neo</sup> Mouse Model

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:

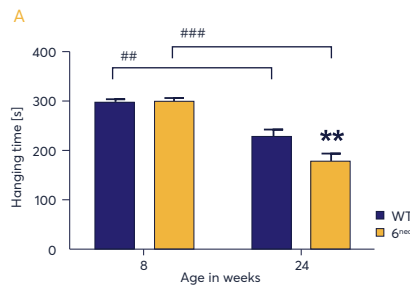
- Reduced body weight
- Astrocytosis

#### Figure 1:

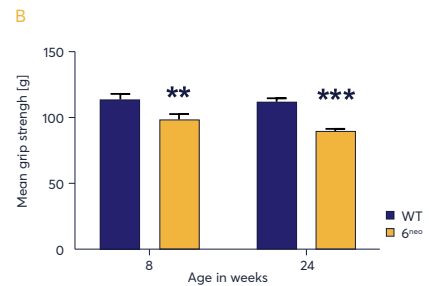
Muscle and motor deficits in the 6<sup>neo</sup> mouse model of Pompe disease. 6<sup>neo</sup> mice at the age of 8 and 24 weeks were tested for hanging time in the wire hanging test (A), for mean grip strength in the grip strength test (B) and for number of slips (C) and slips per speed (D) in the beam walk test (C+D). WT: n = 24; 6<sup>neo</sup>: 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.

Figure 1

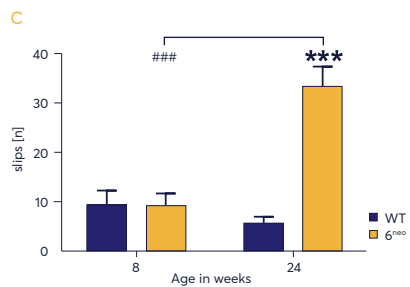
### Wire Hanging



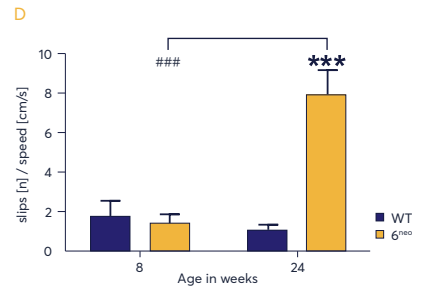
### Grip Strength



### Beam Walk



### Beam Walk



#### 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.

#### Scantox

Discovery

#### Important note:

Representative data are shown throughout this document. However, biological variability might cause deviations from shown data.

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