

## Sanfilippo Syndrome



#### Figure 1:

Behavioral analyses of MPS IIIA mice compared to wild type (WT) littermates in the three chamber social test and Barnes maze test.

- **A:** Social approach in the social interaction test.
- **B:** Time animals spent in the target quadrant during the probe trial of the Barnes maze test at the age of 30 weeks.
- **A:** Two-Way ANOVA with Bonferroni's post hoc test.
- **B:** One Sample t-test; significances label differences compared to 25% (dotted line); \*p<0.05; \*\*p<0.01; \*\*\*p<0.0001.

#### Figure 2:

Histological analysis of MPSIIIA mice compared to WT littermates for neuroinflammation in the cortex.

- A: Immunoreactive area (IR) in percent of GFAP labeling. Mean+SEM; unpaired t-test. \*\*\*p<0.001.
- **B:** Representative images of GFAP labeling in the cortex of MPSIIIA and WT animals at the age of 30 weeks.

#### Figure 3:

Histological analysis of MPSIIIA mice compared to WT littermates for lysosomal/endosomal membrane alterations in the cortex.

- **A:** Immunoreactive area (IR) in percent of LIMP2 labeling. Mean+SEM; unpaired t-test. \*\*\*p<0.001.
- **B:** Representative images of LIMP2 labeling in the cortex of MPSIIIA and WT animals at the age of 30 weeks.

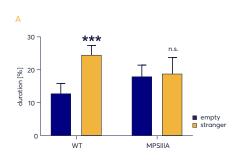
#### MPSIIIA Mouse Model

MPS IIIA mice (JAX#003780) contain a spontaneous Sgsh mutation, resulting in an almost complete loss of N-sulfoglucosamine sulfohydrolase activity. Mice show typical pathological features of the Sanfilippo syndrome A:

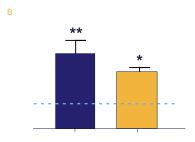
- · Cognitive deficits
- · Social behavior deficits

- Neuroinflammation
- Lysosomal pathology

### Figure 1 Social Interaction Approach



#### **Probe Trial**

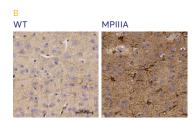


#### Figure 2

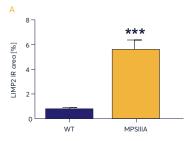
**Astrogliosis** 

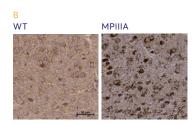
# 6 CPAP II due of [%]

MPSIIIA



Lysosomal and Endosomal Membranes





#### Scantox

Discovery

#### Important note:

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

#### Scantox Group, HQ

Hestehavevej 36A, Ejby DK – 4623 Lille Skensved clientservice@scantox.com www.scantox.com +45 5686 1500

#### © Scantox A/S

Scantox is a registered trademark of Scantox A/S.

