



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

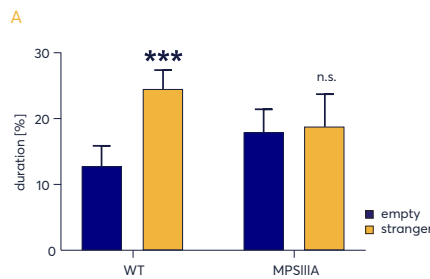
**A:** Social approach in the social interaction test.

**B:** Time animals spent in the target quadrant during the probe trial of the Barnes maze test at the age of 30 weeks.

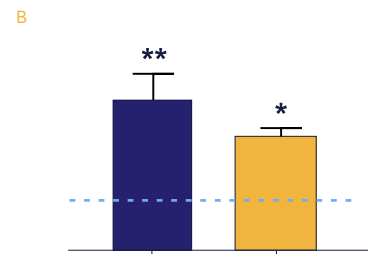
**A:** Two-Way ANOVA with Bonferroni's *post hoc* test.

**B:** One Sample t-test; significances label differences compared to 25% (dotted line); \* $p < 0.05$ ; \*\* $p < 0.01$ ; \*\*\* $p < 0.0001$ .

**Figure 1**  
**Social Interaction Approach**



**Probe Trial**

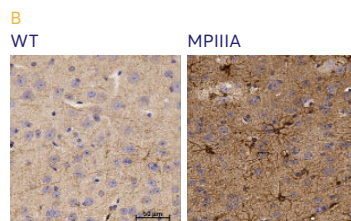
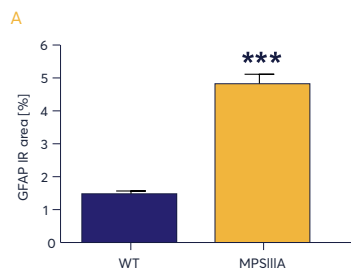


**Figure 2:** Histological analysis of MPSIIIA mice compared to WT littermates for neuroinflammation in the cortex.

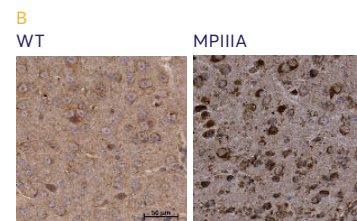
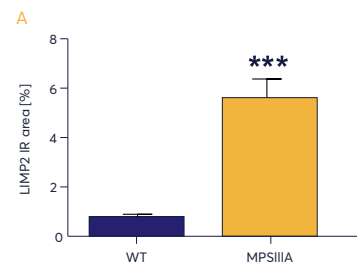
**A:** Immunoreactive area (IR) in percent of GFAP labeling. Mean+SEM; unpaired t-test. \*\*\* $p < 0.001$ .

**B:** Representative images of GFAP labeling in the cortex of MPSIIIA and WT animals at the age of 30 weeks.

**Figure 2**  
**Astrogliosis**



**Figure 3**  
**Lysosomal and Endosomal Membranes**



**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.

