scantox

In vivo Animal Models

Sanfilippo Syndrome

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 wildtype (WT) littermates in the Three Chamber social test and Barnes maze test.

A: social approach in the social interaction test and (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: E: 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.

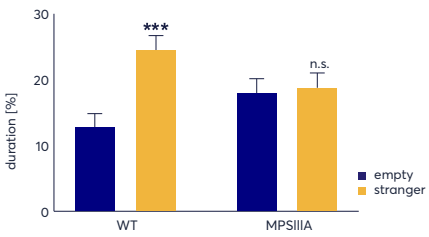
Figure 3: Histological analysis of MPSIIIA mice compared to WT littermates for lysosomal/endosomal membrane alterations in the cortex.

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.

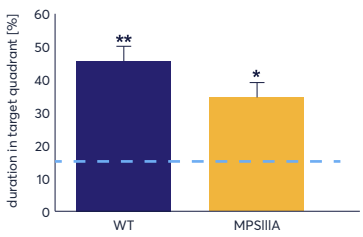
Social Interaction Approach

Figure 1: A



Probe Trial

Figure 1: B



Astrogliosis

Figure 2: A

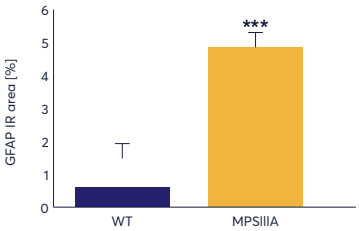


Figure 2: B WT

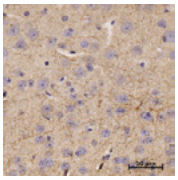
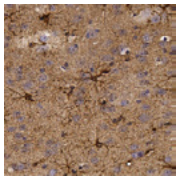


Figure 2: B MPIIIA



Lysosomal and Endosomal Membranes

Figure 3: A

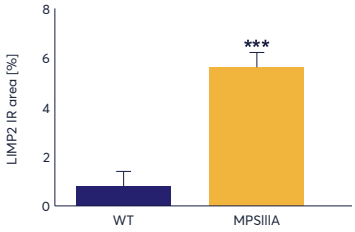


Figure 3: B WT

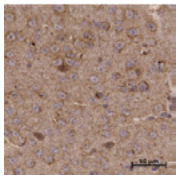
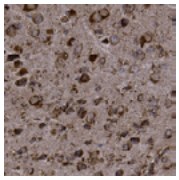


Figure 3: B MPIIIA



Scantox
Discovery

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

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