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In vivo Animal Models

Gaucher Disease

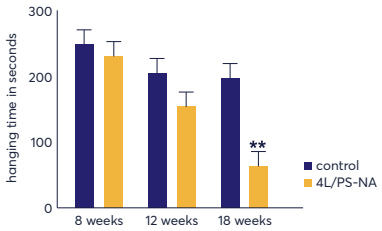
4L/PS-NA Mouse Model

4L/PS-NA mice express low levels of prosaposin and saposins with an additional point mutation in V394L/V394L of glucocerebrosidase. Mice show typical pathological features of the Gaucher disease:

- Motor deficits
- Increased GlcCer and GlcSph substrate levels in brain
- Neuroinflammation
- Visceral pathology in spleen, liver, lung, thymus

Wire Suspension

Figure 1:



RotaRod

Figure 1:

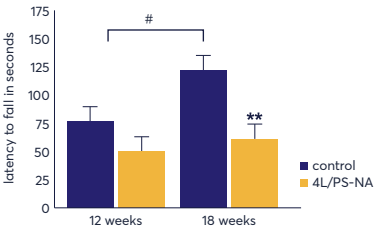
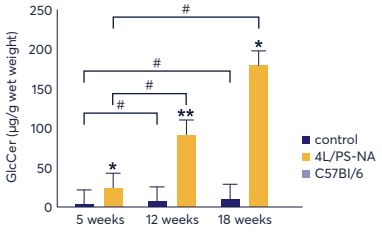


Figure 1:
Wire suspension and RotaRod test of 4L/PS-NA mice over age. Wire suspension time in seconds and latency to fall off the rotating rod of 4L/PS-NA over age. n = 7 per group; Two-way ANOVA with Bonferroni's post hoc test ; Mean + SEM; *p<0.05, **p<0.01.

Glucosylceramide

Figure 2:



Glucosylsphiningosine

Figure 2:

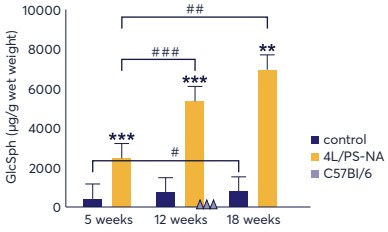
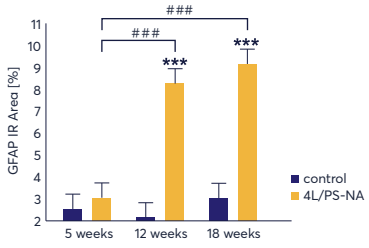


Figure 2:
Quantification of glucosylceramide and glucosylsphingosin in 4L/PS-NA mice over age. Whole brain homogenates of 5, 12 and 18 week old 4L/PS-NA mice, control animals as well as C57Bl/6 mice were analyzed for glucosylceramide and glucosylsphingosin levels. Two-way ANOVA with Bonferroni's post hoc test. n = 3 - 5. Mean + SEM. *p<0.05; **p<0.01; ***p<0.001.

Astrocytosis

Figure 3: A



Astrocytosis

Figure 3: B

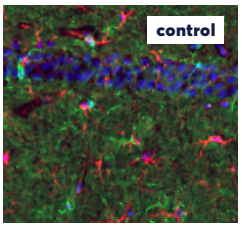


Figure 3: B

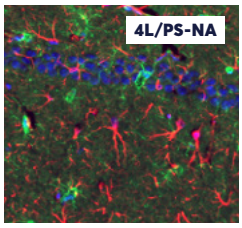


Figure 3:
Cortical astrocytosis and activated microglia of 4L/PS-NA mice over age.
A: Quantification of GFAP immunoreactive (IR) area in percent at the age of 5, 12 and 18 week. n = 5; Two-way ANOVA with Bonferroni's post hoc test. Mean + SEM. ***p<0.001.
B: Representative images of GFAP, IBA1 (activated microglia) and DAPI labeling in 18 week old 4L/PS-NA mice.

Sun Y, Quinn B, Witte DP, Grabowski GA. Gaucher disease mouse models: point mutations at the acid beta-glucosidase locus combined with low-level prosaposin expression lead to disease variants. J Lipid Res. 2005 Oct;46(10):2102-13.

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