In vivo Animal Models

Niemann-Pick Disease

NPC1-/- Mouse Model

Mice homozygous for the recessive NIH allele of the Niemann Pick type C1 gene (Npc1m^{1}N) show a dual deficiency of sphingomyelinase and glucocerebrosidase activity (JAX# 003092). Animals are bred on a BALB/c OlaHsd background. These mice show typical pathological features of the Niemann-Pick disease.

- Increased cholesterol levels in liver
- Lipid accumulation in cerebellum and hippocampus
- Progressive neuronal loss
- Neuroinflammation
- Altered APP expression and accumulation of amyloid-β peptides
- Phosphorylated tau
- Motor coordination deficits

Figure 1:

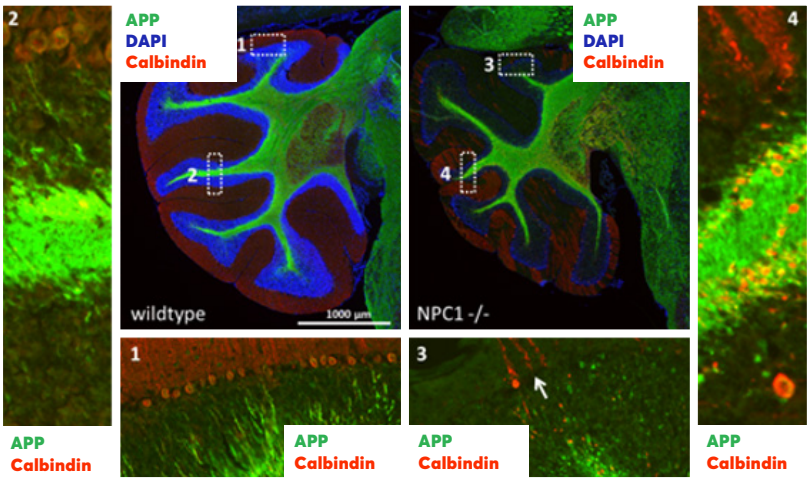

Figure 1:
Neuron loss and neuroinflammation in the cerebellum, including Purkinje cells. In anterior parts of the cerebellum the Calbindin immunoreactive area (red) Purkinje cell network breaks down and only small stripes remain as indicated by the arrow (3). Healthy Purkinje cells are positive for APP (green, Y188). Nuclei are labeled with DAPI (blue).

RotaRod

Figure 2:

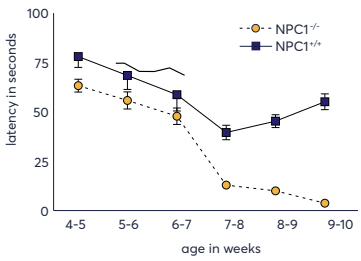

Figure 2:
RotaRod test of 4 to 10 weeks old NPC1-/- mice. Longitudinal characterization of latency to fall off the rod of NPC1-/- and wild type control animals NPC1+/+. n = 36 per group; Mean ± SEM. Two-way ANOVA with Bonferroni's post hoc test. ***p<0.001.

Santiago-Mujica E, Flunkert S, Rabl R, Neddens J, Loeffler T, Hutter-Paier B. Hepatic and neuronal phenotype of NPC1^{-/-} mice. Heliyon. 2019 Mar 14;5(3):e01293.

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