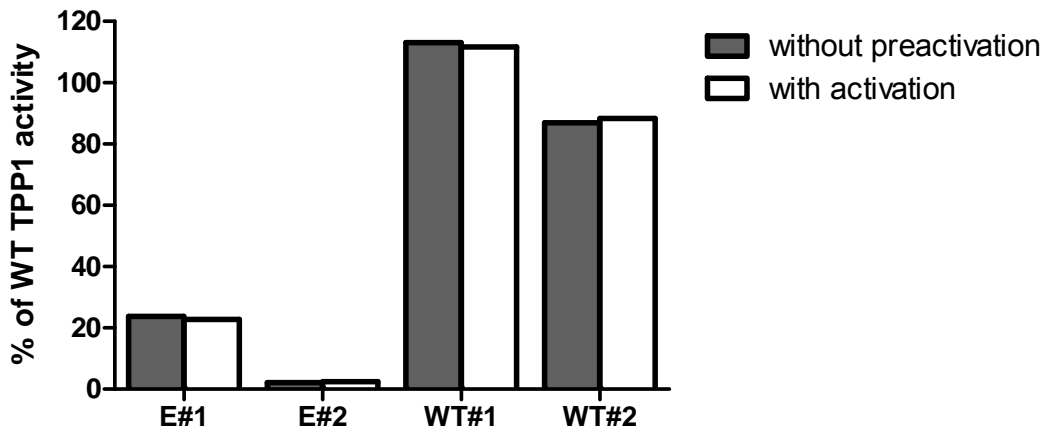


Supplemental Figure S1 TPP1 proenzyme is activated after delivery into brain.

(a) fifteen-week old *Tpp1*^{-/-} mice (n=2) were administered a single dose of 20 μ L 7.2 μ g/ μ L rhTPP1 and killed 48 hours afterwards. Age matched wildtype mice (n=2) were included as control. TPP1 activity in brain is measured with (open bar) or without pre-activation (grey bar) and normalized to the wildtype control. **(b)** fifteen-week old *Tpp1*^{-/-} mice were administered 3 daily dose of artificial CSF (V, n=4) or 80 μ L 10 μ g/ μ L rhTPP1 (E, n=6). After 48 hours, mice were killed and TPP1 was detected in brain homogenates by western blotting using a rabbit polyclonal anti-TPP1 antibody. Samples were normalized to protein amount (7 μ g per lane) and an age matched wildtype mouse was included as control (wt). Positions of *in vitro* activated rhTPP1 (act) and non-activated proTPP1 (pro) were also shown. Note that there is a small difference in the migration of TPP1 activated *in vitro* and *in vivo*, which probably reflects the processing of N-linked glycans within the lysosome.

a



b

