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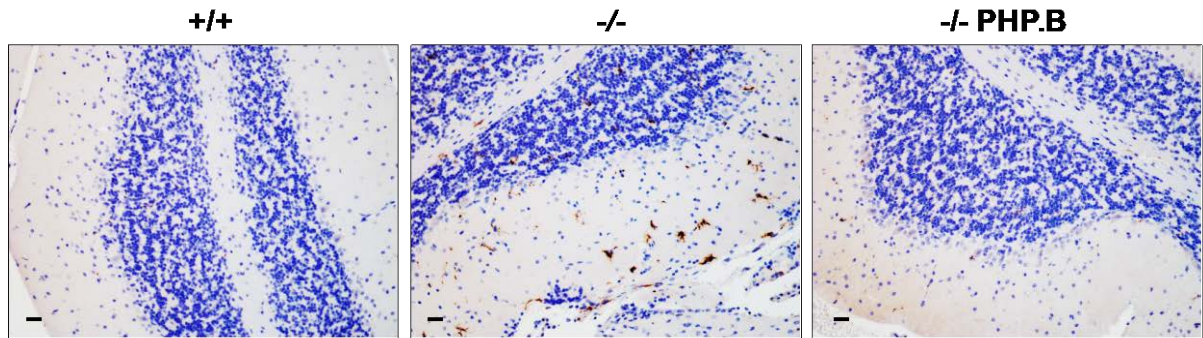
Supplemental Information

A Single Intravenous Injection of AAV-PHP.B-*hNDUFS4* Ameliorates the Phenotype of *Ndufs4*^{-/-} Mice

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Supplemental Material

Supplemental Figure 1. Scattered CD68-positive cells in the posterior cerebellar posterior lobule. Scale bars, 10 μ m.

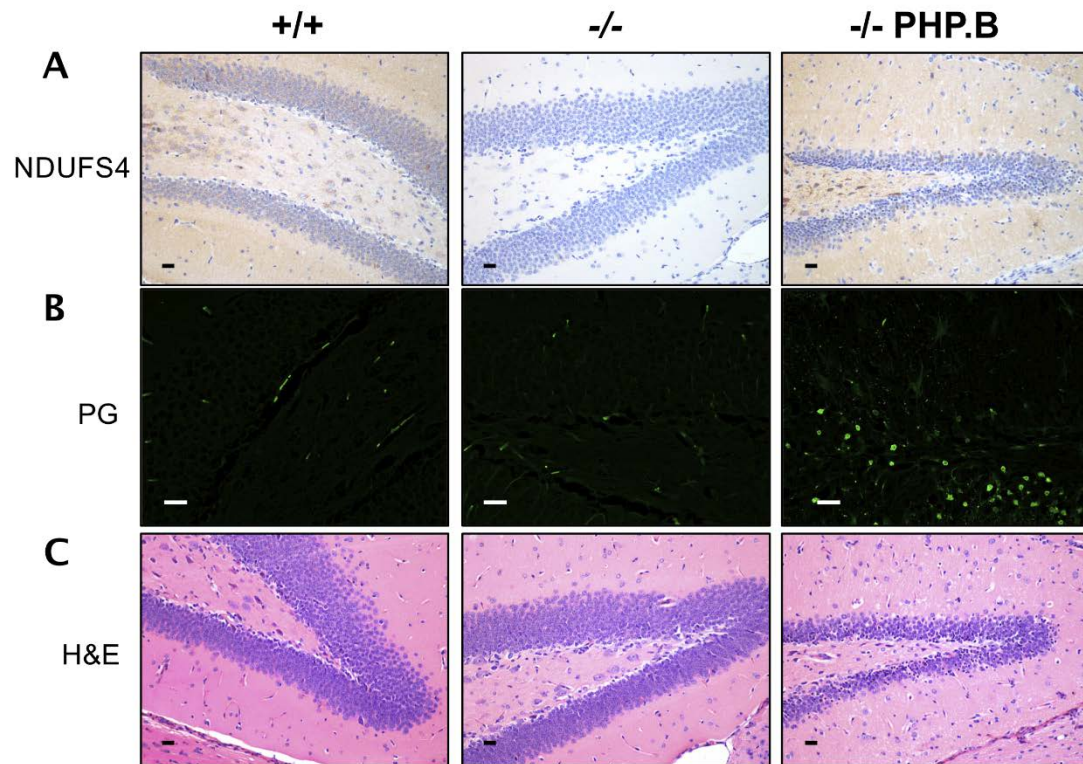


Supplemental Figure 2. Characterization of the dentate gyrus of the hippocampus.

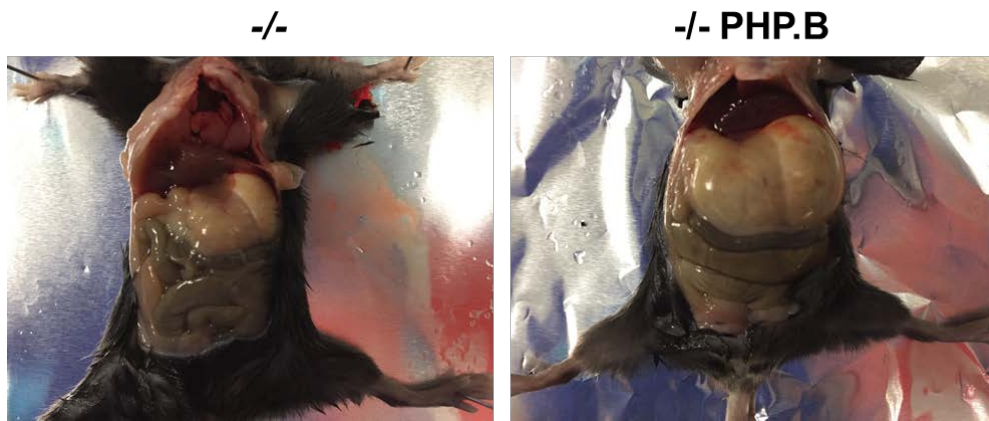
A) Immunodecoration with antiNDUFS4 antibody. Scale bars, 10 μm .

B) PathoGreen staining. Scale bars, 20 μm .

C) H&E staining. Scale bars, 10 μm .



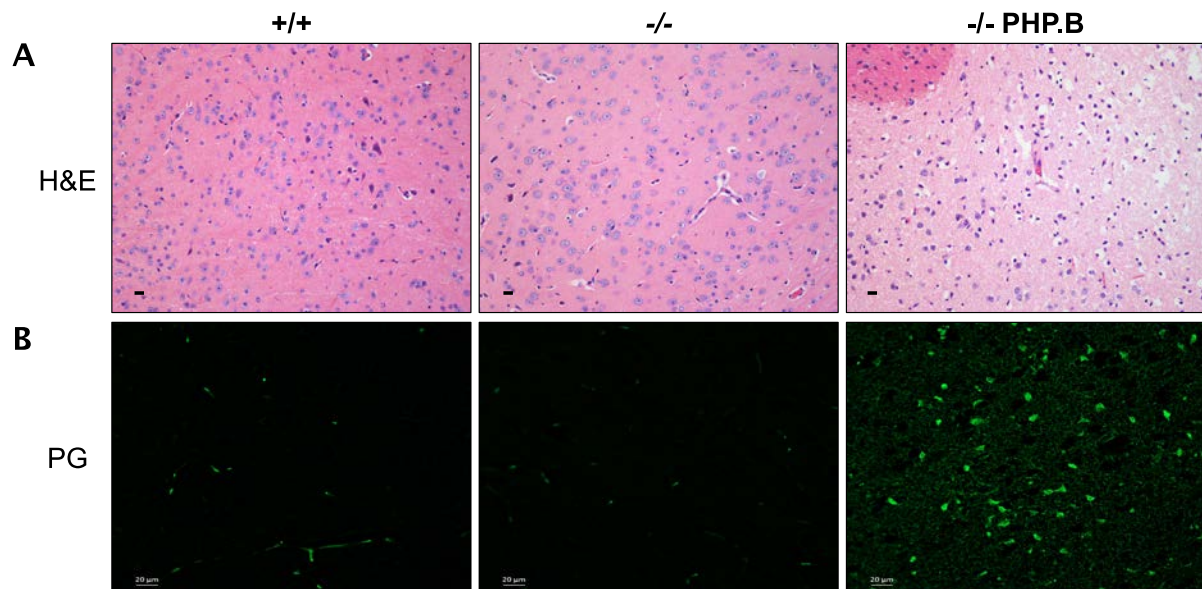
Supplemental Figure 3. Massively enlarged stomach of AAV-PHP.B-*hNDUFS4* mice.



Supplemental Figure 4. Massive neurodegeneration in the pre-optic area of a *Ndufs4*^{-/-} mouse sacrificed because of poor clinical conditions.

A) H&E staining. Scale bars, 10 μ m.

B) PathoGreen staining. Scale bars, 20 μ m.



Supplemental Figure 5. Western-blot immunovisualisation of LY6A protein in brain homogenates of newborn (P1) and P30 mice. CBB: Coomassie brilliant blue staining; β -tubulin was used as an internal standard for protein loading.

