Supplementary Material

Legends for Supplementary Figures:

Figure S1: Oxidative stress and disrupted energy metabolism in Ppt1-KO mice and INCL cells

(A) Western blot analysis of SOD2 protein levels from the cerebral cortex of 1-, 3- and 6-month old WT mice and their *Ppt1*-KO littermates. (**B**) Western blot analysis of p-AMPK and PGC-1α protein levels in the INCL patient fibroblast with AICAR treatment for various time periods. ß-Actin was used as loading control. (**C**) Western blot analysis of FoxO3a protein levels from the cerebral cortex of 1-, 3- and 6-month old WT mice and their *Ppt1*-KO littermates. **(D)** Western blot analysis of FoxO3a protein levels in the brain tissue lysates of WT mice and their *Ppt1*-KO littermates on normal diet, or on RSV diet.

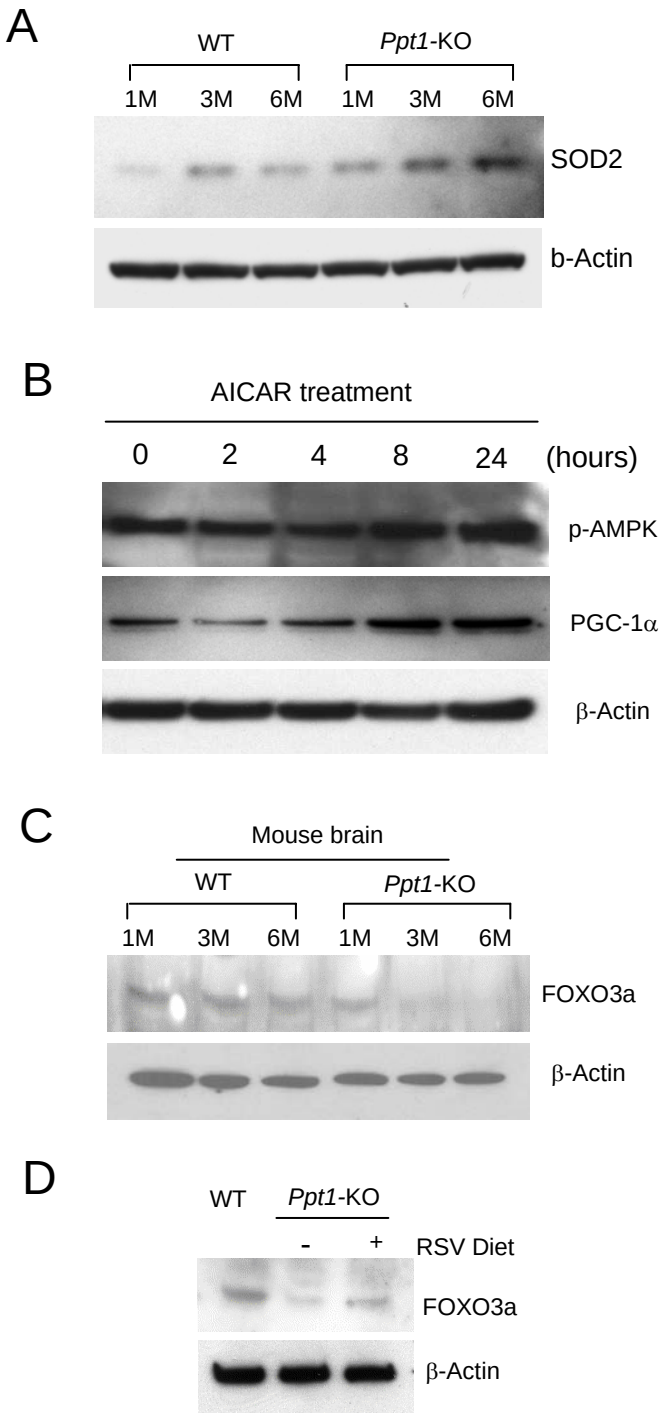
Figure S2: Resveratrol stimulates mRNA levels of transcription factors regulating mitochondrial biogenesis.

(A) A comparison of Mfn1-, Fis1- and TFAM-mRNA levels in untreated normal and INCL fibroblasts determined by real-time RT-PCR. (**B**) Levels of Mfn1-, Fis1- and TFAM-mRNAs in untreated and RSV (20μM)-treated cultured INCL fibroblasts. Note that RSV treatment increases mRNA levels of all 3 transcription factors in a time-dependent manner. (**C**) Stimulation of Mfn1-, Fis1- and TFAM-mRNA levels in brain tissues of WT mice and *Ppt1*-KO littermates by normal and RSV-diets. Results are expressed as the mean ± SEM. (n=3). Asterisks indicate levels of significance (*, $p<0.05$; **, $p<0.01$).

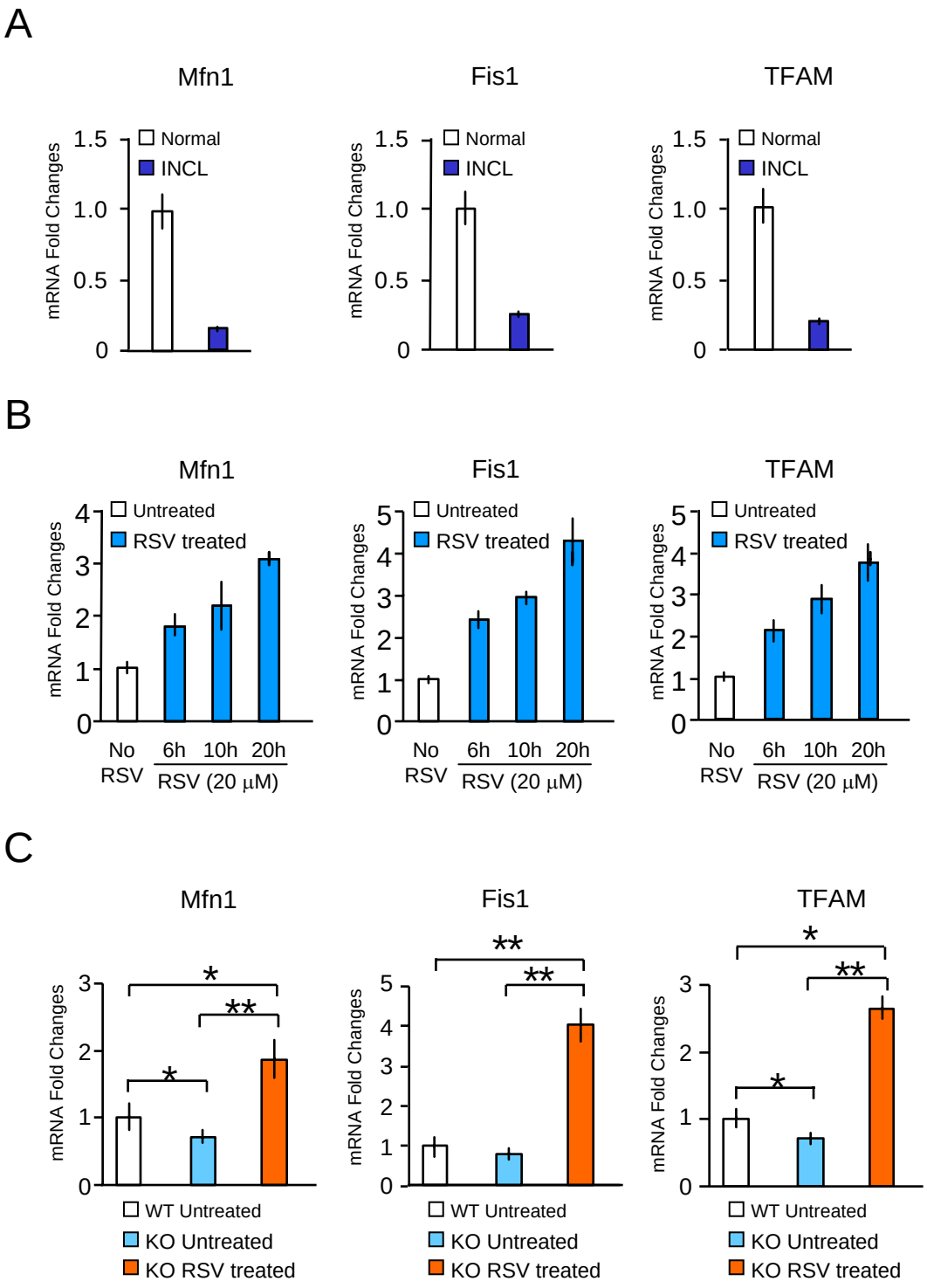
Figure S3: **Resveratrol down-regulates PI3- and Akt-kinases in *Ppt1*-KO mouse brain.**

(**A**) Western blot analysis of PI3K in cerebral cortex of 1-, 3- and 6-month-old WT mice and their *Ppt1*-KO littermates. (**B**) Western blot analysis of p-Akt (Ser 473), p-AKT (Thr 308) and control Akt proteins in cerebral cortex of 1-, 3- and 6-month-old WT mice and their *Ppt1*-KO littermates. (**C**) Western blot analysis of p-Akt (Ser 473), p-Akt (Thr 308), control Akt proteins and PI3K in cerebral cortex of *Ppt1*-KO mice on normal- or RSV-diet. ß-actin was used as loading control.

Supplementary Figure 1



Supplementary Figure 2



Supplementary Figure 3