

## Supplemental Figures:

### SFig.1. Abnormal behavior of Cer2<sup>gt/gt</sup> mice (Video)

In contrast to wild type and heterozygous litter mates, the CerS2<sup>gt/gt</sup> mouse (marked by x in the initial frames of the movie) has difficulties to start motor activity, in the absence of obvious spasms or muscular defects, collectively suggesting a neurological malfunction of the motor system

SFig.2. Expression of CerS2 promoter-driven  $\beta$ -galactosidase activity in the kidney of CerS2<sup>gt/gt</sup> mice. **(A and B)** Staining is most intense in the medullary region, there being concentrated on collecting ducts. In the renal cortex, glomeruli (G in **B**) are largely negative, while most tubules exhibit a low staining intensity. Only proximal tubules (PT in **B**) identified by both their contact to renal capsule and their connection to the Bowman capsules of glomeruli (arrow in **B**) featured an intermediate staining density. **(C - F)** Histologically, the kidneys of CerS2<sup>gt/gt</sup> mice up to 9 months of age appeared largely normal. The only visible effect is the occurrence of several clefts within the renal tissue (arrows) mainly at the corticomedullary junction, often apposed to larger veins. **(E)** represents the enlarged box of **D**. These spaces (arrows in **D** to **F**) were lined by an incomplete layer of flat cells, with glomeruli and tubules sometimes protruding into their lumen (**F**). Calibration bars correspond to 1 mm in **A**, 200  $\mu$ m in **B**, 500  $\mu$ m in **C**, 1 mm in **D**, 150  $\mu$ m in **E** and **F**.

Supplemental Figure 1 (Video)



Supplemental Figure 2

