



**Fig. S1. Effect of copper-deficient diet on body weight of *Npc1*<sup>+/+</sup> and *Npc1*<sup>-/-</sup> mice, and on survival of *Npc1*<sup>-/-</sup> mice.** Body weight (A) of *Npc1*<sup>+/+</sup> (white symbols) and *Npc1*<sup>-/-</sup> (black symbols) mice fed with copper adequate (CuA) or copper-deficient diet (CuD) and survival (B) of *Npc1*<sup>-/-</sup>. Body weight data represent mean  $\pm$  SEM. At least five animals per group were analyzed.