

Figure S1

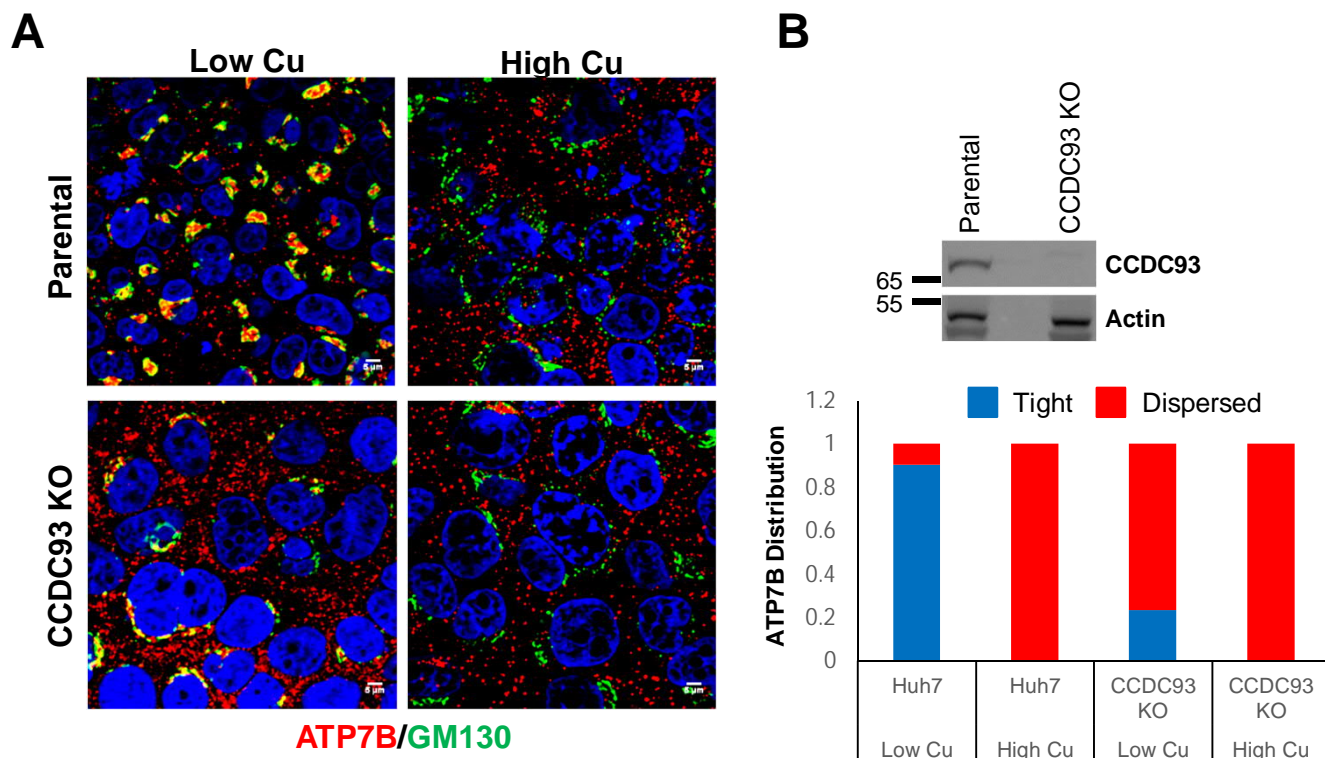


Figure S1: Depletion of CCDC93 impairs ATP7B trafficking. (A) Huh-7 parental cells and a CCDC93 knockout sub-line were exposed to high or low copper-containing media. Following this, cells were immunostained for ATP7B (red) and the Golgi marker GM130 (green). A nuclear dye was also used (blue). Immunofluorescence images are shown. Scale bar, 5 μ m. (B) Immunoblot for CCDC93 and actin (top panel) and quantification of ATP7B distribution pattern in the indicated cell lines.

Figure S2

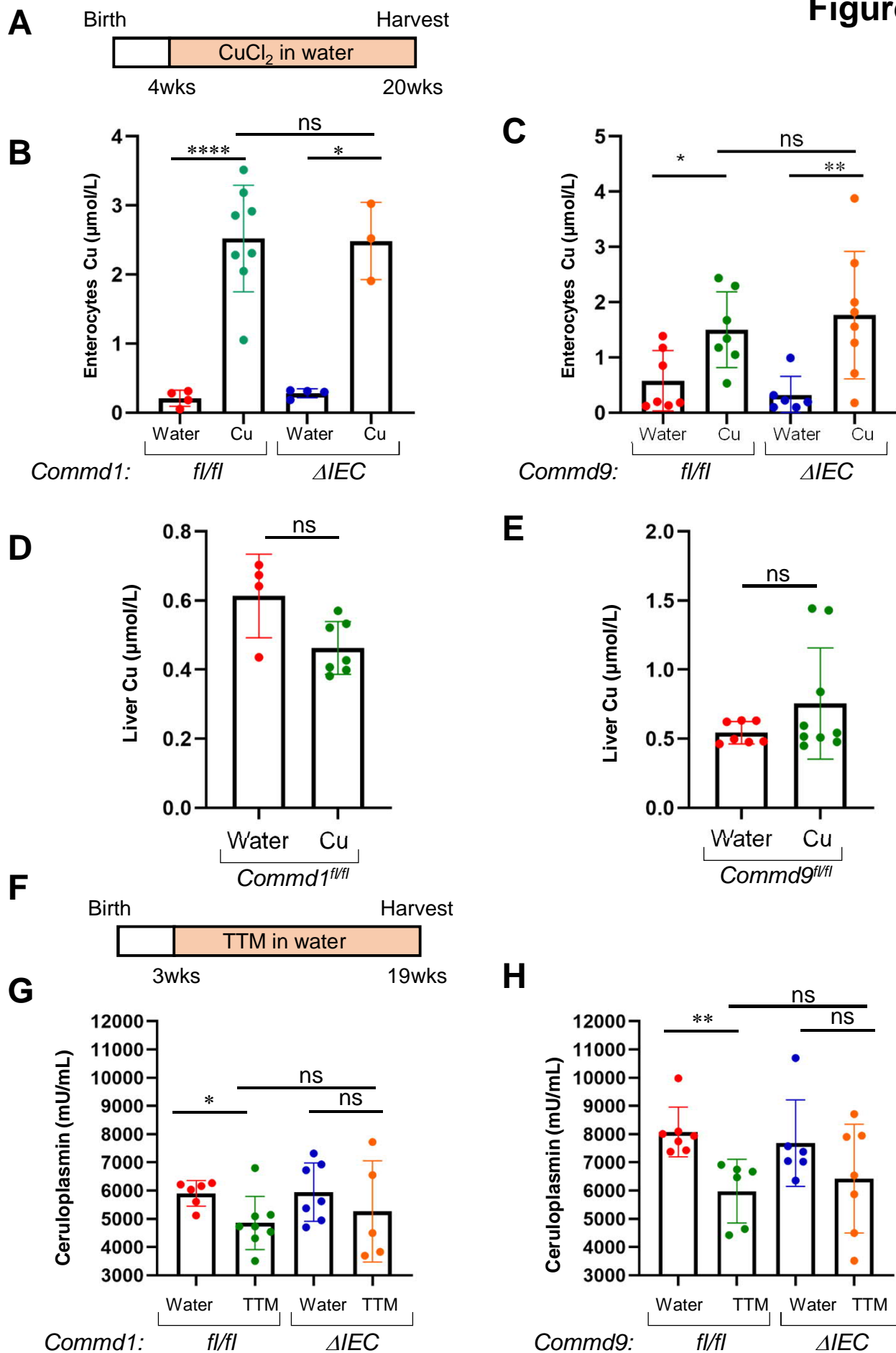


Figure S2: Enteric *Commd* knockout does not impair copper balance. (A) Schematic depicting copper treatment course. (B) Enterocyte copper concentrations ($\mu\text{mol/L}$) were measured in isolated enterocytes of *Commd1^{ΔIEC}* mice (water control: n=4, copper treated: n=3) and corresponding floxed control animals (water control: n=4, copper treated: n=8) at the conclusion of the high copper diet treatment depicted in (A). Results for individual mice are plotted along with the mean and s.e.m. for each group; * $P < 0.05$, **** $P < 0.0001$ and ns, not significant (unpaired two-tailed t test). (C) Same analysis as in (B) but for *Commd9^{ΔIEC}* (water control: n=6, copper treated: n=8) the corresponding littermate controls (water control: n=7, copper treated: n=7). * $P < 0.05$, ** $P < 0.01$ and ns, not significant (unpaired two-tailed t test) (D) Hepatic copper concentrations were measured in dried liver tissue of *Commd1^{fl/fl}* animals on a water (n=4) or high copper diet (n=7). Results for individual mice are plotted along with the mean and s.e.m. for each group; ns, not significant (unpaired two-tailed t test). (E) Same analysis as in (D) but for *Commd9^{fl/fl}* animals on a water (n=7) or high copper diet (n=9). (F) Schematic depicting tetrathiomolybdate (TTM) treatment course. (G) Ceruloplasmin levels (mU/mL) were measured in plasma of *Commd1^{ΔIEC}* mice (water control: n=7, TTM treated: n=5) and corresponding floxed control animals (water control: n=6, TTM treated: n=8) on TTM or control (water) diets. Results for individual mice are plotted along with the mean and s.e.m. for each group; * $P < 0.05$, and ns, not significant (unpaired two-tailed t test). (H) Same analysis as in (G) but for *Commd9^{ΔIEC}* mice (water control: n=6, TTM treated: n=7) and corresponding floxed animals (water control: n=7, TTM treated: n=6). ** $P < 0.01$ and ns, not significant (unpaired two-tailed t test).