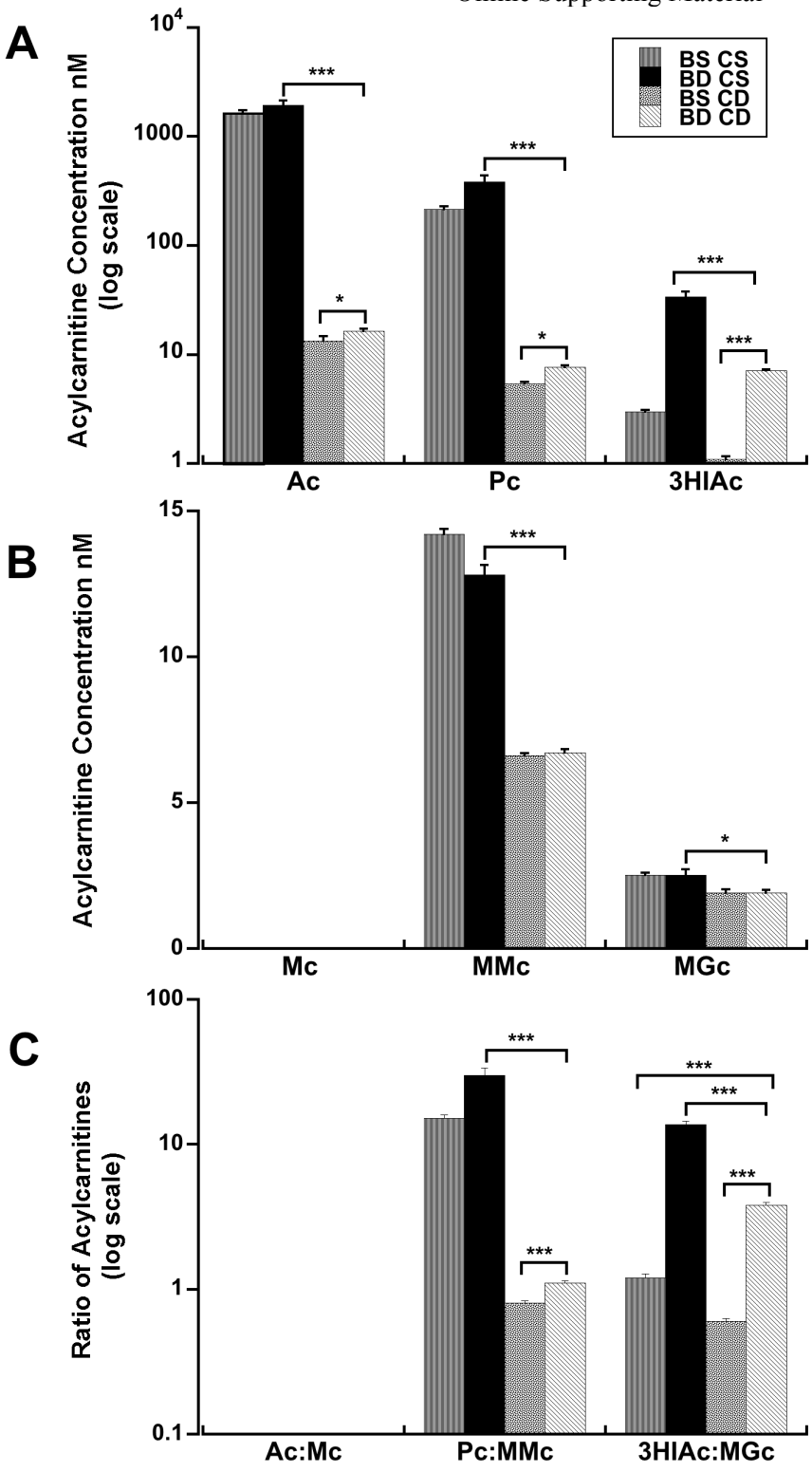

Supplemental Figure 4. Effect of combined biotin deficiency and carnitine deficiency on **extracellular** acylcarnitine concentrations and acylcarnitine ratios at the point of greatest depletion of biotin, carnitine, or both in HepG2 cells. A. Acylcarnitines Ac, Pc, and 3HIAc that arise from the acyl-CoAs that are substrates of the biotin-dependent carboxylases. B. Acylcarnitines Mc, MMc, and MGc that arise from the acyl-CoAs that are products of the biotin-dependent carboxylases. C. Substrate:product ratios of the acylcarnitines. Values are mean ± SD, n=4. Student's t test**:** * $P < 0.05$, *** $P < 0.001$. BD CD, biotin deficient carnitine deficient; BD CS, biotin deficient carnitine sufficient; BS CD, biotin sufficient carnitine deficient; BS CS, biotin sufficient carnitine sufficient; Ac, acetylcarnitne; 3HIAc, 3-hydroxyisovalerylcarnitine; Mc, malonylcarnitine; MGc, 3-methylglutarylcarnitine; MMc, methylmalonylcarnitine; Pc, propionylcarnitine.