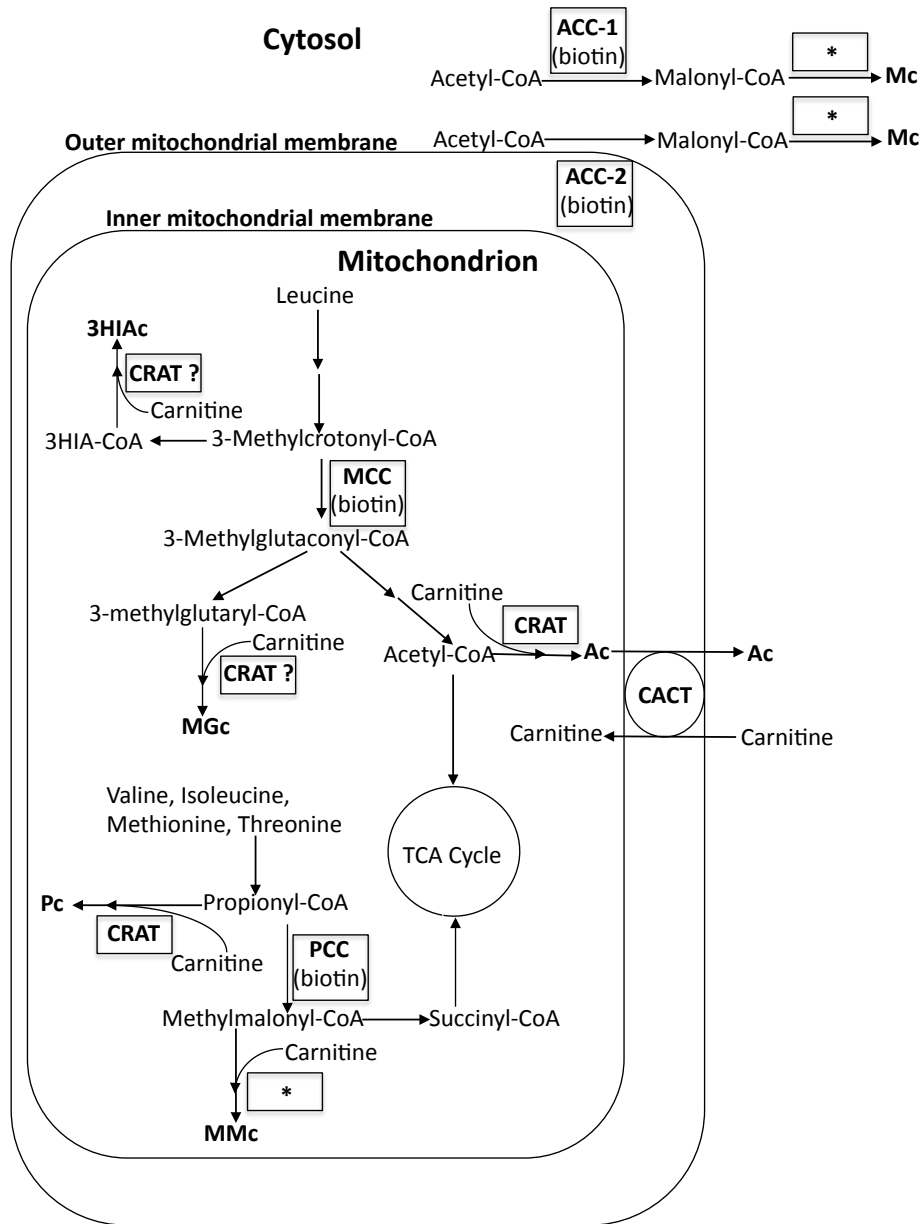


Online Supporting Material



Supplemental Figure 1. Metabolic activities of four carboxylases involved in formation of the acyl-CoAs and carnitine-dependent transport of acyl groups through mitochondrial membrane via CACT. Transport is depicted only for Ac; transport out of mitochondria of Pc, MMc, 3HIAc and MGc are also mediated by CACT. * and ?: Enzyme and/or subcellular sites of Mc, MMc, 3HIAc, and MGc formation are unknown. ACC-1, acetyl-CoA carboxylase 1; ACC-2, acetyl-CoA carboxylase 2; Ac, Acetylcarnitine; CACT, carnitine acylcarnitine translocase; CRAT, carnitine acetyltransferase; 3HIAc, 3-hydroxyisovalerylcarnitine; Mc, malonylcarnitine; MCC, methylcrotonyl-CoA carboxylase; MGc, 3-methylglutaryl carnitine; MMc, methylmalonylcarnitine; Pc, propionyl carnitine; PCC, propionyl-CoA carboxylase.