CPA2 encodes the large subunit of carbamoyl-phosphate synthase, a cytosolic enzyme in the arginine biosynthetic pathway. The small subunit, encoded by CPA1, binds and cleaves glutamine, while the large subunit, Cpa2p, binds the remaining substrates and carries out all other reactions catalyzed by the heterodimer. Interaction between Cpa1p and Cpa2p increases the Vmax of Cpa2p, perhaps by causing a conformational change in Cpa2p. Yeast carbamoyl-phosphate synthase shares sequence similarity with heterodimeric and single subunit carbamoyl-phosphate synthases from E. coli and eukaryotes including rat, human, and hamster. In humans, carbamoyl-phosphate synthase deficiency causes hyperammonemia.