CPA1 encodes the small subunit of carbamoyl-phosphate synthase, a cytosolic enzyme in the arginine biosynthetic pathway. The small subunit, Cpa1p, binds and cleaves glutamine, while the large subunit, encoded by CPA2, 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. Both transcription and translation of CPA1 are repressed in the presence of arginine; the translational repression requires sequences within a small ORFupstream of the coding sequence in the CPA1 mRNA. 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.