ARG3 encodes ornithine carbamoyltransferase, a cytosolic enzyme involved in the biosynthesis of arginine from ornithine. Like other genes encoding arginine biosynthetic enzymes, ARG3 is transcriptionally repressed in the presence of arginine and is regulated by general amino acid control. Arginine-responsive transcription factors, including Arg80p, Arg81p, Arg82p, and Mcm1p, have been identified, as have their target upstream activating sequences in ARG3. Arg3p is related to ornithine carbamoyltransferases from E. coli, Aspergillus nidulans, rat, and human. Ornithine carbamoyltransferase deficiency causes hyperammonemia in humans.