ARG8 encodes acetylornithine aminotransferase, a mitochondrial matrix enzyme that catalyzes the fourth step in the biosynthesis of ornithine, an intermediate in arginine biosynthesis. Arg8p is 68% identical to the acetylornithine aminotransferase from Kluyveromyces lactis, and the K. lactis gene can complement an S. cerevisiae arg8 mutant. Arg8p is also similar to E. coli ArgD. Like other genes encoding arginine biosynthetic enzymes, ARG8 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, and their target upstream activating sequences in ARG8, have been identified. The ARG8 sequence has been recoded using the mitochondrial genetic code, for use as a marker for mitochondrial transformation. A construct in which the recoded ARG8 replaces the COX3 coding sequence complements a deletion of the nuclear ARG8 gene, and its expression requires COX3 mRNA-specific translational activators.