ARG1 encodes argininosuccinate synthase, which catalyzes the eighth step in arginine biosynthesis. Arg1p also plays a catabolic role in the utilization of citrulline as a nitrogen source. Arg1p and other enzymes that catalyze steps in the part of the arginine biosynthetic pathway going from ornithine to arginine are cytosolic. Like other genes encoding arginine biosynthetic enzymes, ARG1 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 ARG1. Arg1p is similar to the E. coli argininosuccinate synthase, and similar enzymes have been identifed in archaebacteria and mammals. Mutations in the human gene encoding argininosuccinate synthase cause citrullinemia.