ORT1 encodes an ornithine transporter localized to the mitochondrial inner membrane. In yeast, ornithine is synthesized in the mitochondrial matrix and transported to the cytosol, where it is converted to arginine. Mutations in ORT1 cause slow growth in the absence of arginine; the residual growth may be due to Bac1p, another mitochondrial inner membrane transporter. Mutations in ORNT1, a human gene orthologous to ORT1, are associated with hyperammonemia-hyperornithinemia-homocitrullinuriasyndrome.