ADE13 encodes adenylosuccinate lyase, which is involved in catalysis of two reactions in the purine nucleotide biosynthetic pathway: the first in the de novo pathway leading to formation of inosine monophosphate, and the second in the interconversion of IMP to adenosine monophosphate.Expression of ADE13 and most of the other purine biosynthetic genes is repressed by adenine and activated by transcription factors Bas1p and Pho2p. Reduction- or loss-of-function mutations in ADE13 relieve the transcriptional repression of the ADE genes by adenine, suggesting that AMP plays a role in mediating adenine repression. Only the most severe ade13 mutations also cause adenine auxotrophy, indicating that low levels of Ade13p activity are sufficient for growth in the absence of adenine.Ade13p is a widely conserved protein, and orthologs have been described in bacteria, chicken, and humans. Mutations in the human gene ADSL cause adenylosuccinase deficiency, a rare autosomal recessive disease that causes severe mental retardation, and in some cases convulsions, autism, and growth retardation.