AGC1 encodes a mitochondrial inner membrane aspartate/glutamate transporter that contributes to the biosynthesis of nitrogen compounds. Agc1p is orthologous to the human proteins aralar) and citrin). Like the human orthologs, Agc1p can act as an aspartate-glutamate antiporter. In contrast to the human orthologs, Agc1p also functions as a uniporter for both glutamate and aspartate, it does not contain an N-terminal EF-hand Ca++-binding motif, and its transport activity is not Ca++ dependent.In yeast, mutations in AGC1 result in decreased synthesis of valine, ornithine and citrulline, and the inability to grow on acetate or oleic acid. In humans, mutations in SLC25A13 are associated with type II citrullinemiaand neonatal-onset (605814>OMIM