CCC2 encodes a P-type copper-transporting ATPasenecessary for the proper uptake of iron. Ccc2p receives copperionsfrom Atx1pand transports them into a late or post-Golgi compartment where they are acquired by the cell-surface iron transporter Fet3p, although there is also an Atx1p-independent pathway. CCC2 expression is regulated by iron and AFT1.CCC2 was originally identified as a gene whose twofold overexpression suppresses the calcium-sensitive growth phenotype of sur1 mutants. Deletion of CCC2 is not lethalbut causes several deficiencies that can be overcome by supplementing the growth medium with copper or, in some cases, iron. Among these are: defective iron uptake, slow growth on ethanol, and slow growth at neutral or alkaline pH. ccc2 null mutants are deficient at making inositolphosphorylceramide D, possibly due to failure to deliver copper to an unknown enzyme.Ccc2p is homologous to two human genes, called ATP7Aand ATP7B. Mutations in ATP7A have been shown to cause Menkes diseaseand Occipital Horn Syndrome. Mutations in ATP7B have been shown to cause Wilson disease. Both ATP7Aand ATP7Bcan complement deletion of CCC2 in yeast, as can Candida albicans CCC2and CCC2 homologs from C. elegans, Arabidopsis thaliana, Trametes versicolor, and Brassica napus. An interesting splice variant of ATP7B that partially complements deletion of CCC2 is found in rats, lacks the copper-binding and transmembrane domains, and is expressed in the pineal gland at night.