ATP11 encodes a protein required for the assembly of mitochondrial ATP synthase 4. The ATP synthase complex utilizes proton motive force to generate ATP from ADP and Piand consists of two major components, soluble F1 and membrane-bound F0, each of which contains many subunits. The catalytic core of the enzyme resides in the F1 component and consists of a hexamer of alternating alphaand betasubunits. Although Atp11p binds selectively to the beta subunit, it is not a part of the ATP synthase complex. Instead, 0006461>Atp11p binds to isolated beta subunits in order to prevent beta subunit aggregation prior to F1 assembly. It is likely that Atp11p is displaced by the alpha subunit during complex assembly. A second protein, Atp12p, functions in a similar manner to prevent aggregation of the alpha subunit prior to F1 assembly. Deletion of ATP11 leads to aggregation of beta subunits and loss of F1 assembly. Although ATP11 is required for the production of functional ATP synthase, it is not essential for life in yeast. Like deletions in many genes necessary for the function or maintenance of mitochondria, loss of ATP11 leads to a \"petite\" phenotype that is slow-growing and unable to survive on nonfermentable carbon sources.