PIM1 encodes the ATP-dependent Lon protease. It is required for degradation of misfolded or mislocalized proteins or of unassembled subunits in the mitochondrial matrix with the assistance of chaperone proteins such as Hsp70and Hsp78. Note that the mitochondrion contains at least two other ATP-dependent proteases, the m-AAA protease consisting of two subunitsand the i-AAA protease, both of which are involved in proteolysis of mitochondrial inner membrane proteins.Cells with null mutations in pim1 are viable, but have abnormal mitochondrial morphology, lose mitochondrial DNA, and are respiratory deficient, being unable to grow on non-fermentable carbon sources such as glycerol, ethanol, or lactate. In addition to its catalytic activity as a protease, Pim1p may also play a role in the assembly of mitochondrial complexes. Overexpression of protease-defective Pim1p suppresses the respiratory growth and respiratory complex assembly defects of cells lacking either of the two subunits of mitochondrial membrane m-AAA protease, Afg3p and Yta12p, while overexpression of protease-capable but ATPase-defective Pim1p does not, implicating the ATPase activity in a chaperone-like function for respiratory complexes. However, pim1 mutants also have defects in the stability of the intron-containing COX1 and COB pre-mRNAs and in the translation of COX1 mRNA, so it may be that Pim1p is involved in the production of these two subunits, perhaps via proteolytic maturation of an intron-encoded mRNA maturase, rather than in their assembly into respiratory complexes.Lon protease is conserved in bacteria, archaea, and eukaryotic mitochondria and peroxisomes. In cerevisiae, the Lon protease is composed of a ring of seven Pim1p subunits, while in E. coli the ring is composed of six identical subunits. Each subunit contains two conserved domains, a central ATPase domain and a carboxyl-terminal domain containing proteolytic activity, in contrast to the arrangement in two-component ATP-dependent proteases such as the 26S proteasome where the ATPase and proteolytic activities are present in different subunits.