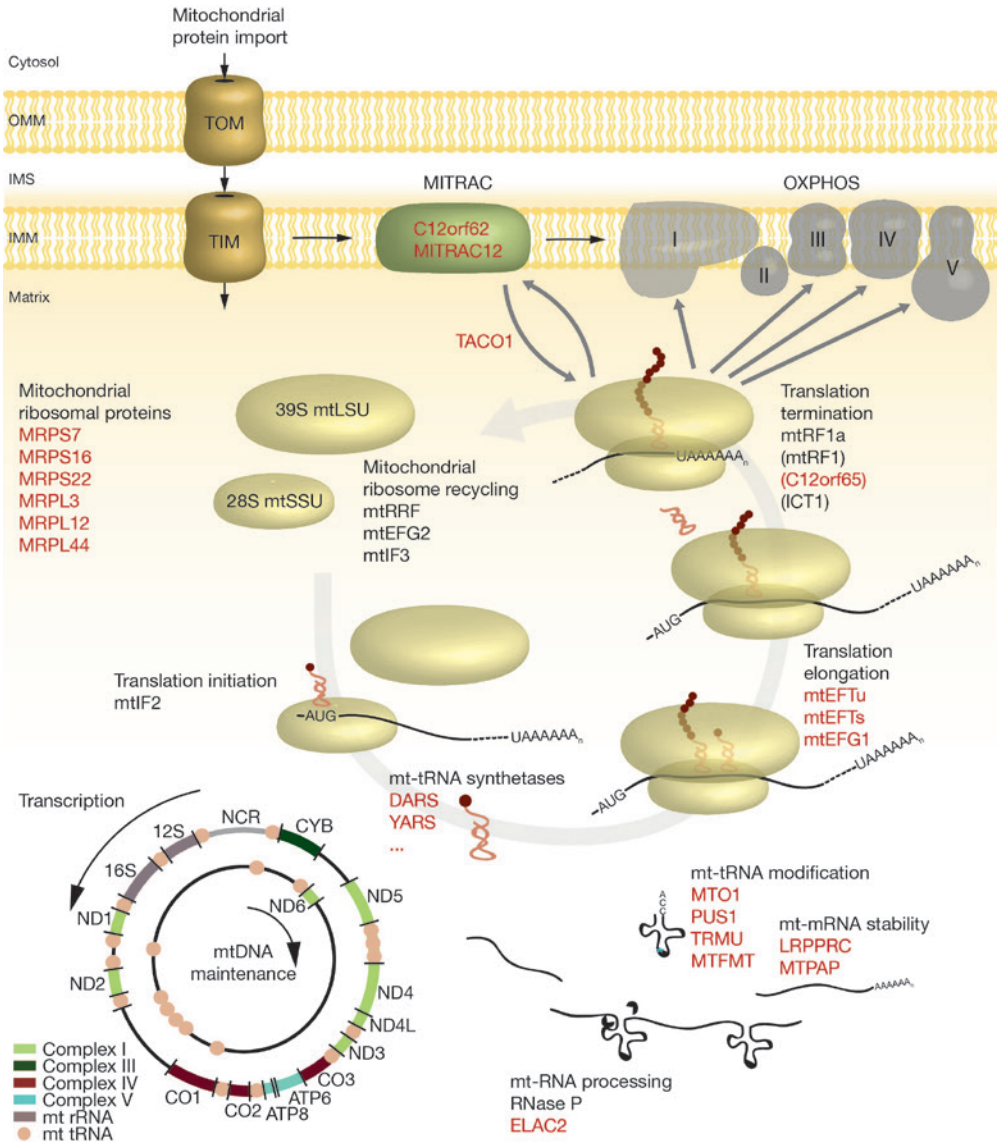

Supplementary infomation S1 (figure) | **Mitochondrial translation disorders.** All components required for the synthesis of mtDNA-encoded proteins need to be imported into the mitochondrial matrix by the mitochondrial protein translocase. Several factors are required for mtDNA replication and transcription1. Many mtDNA mutations have been described to cause isolated or combined complex I, III, IV or V deficiencies. However, there is a growing group of nuclear-encoded factors that are required for the synthesis of mtDNA-encoded proteins, in which mutations were described to cause human disorders (labeled in red). These include factors for RNA processing, mRNA polyadenylation and stability, and tRNA modification enzymes. So far mutations in ten aminoacyl tRNA synthetases have been reported to cause severe disorders. Also the elongation factors mtEFTu, mtEFTs and mtEFG1 are associated with human diseases as well as the mitochondrial class I release factor C12ORF65, and so far six structural subunits of the mitochondrial ribosome. A rather specific defect in the translation of the complex IV core subunit COX1 has been described for mutation in TACO1 and the MITRAC components C12ORF62 and MITRAC12. Factors not labeled in red have so far not been associated with human disorders. Factors in parentheses are class I release factors, for which a role in translation termination is still not clear.

1. Hällberg, B. M. & Larsson, N.-G. Making Proteins in the Powerhouse. *Cell Metabolism* **20**, 226–240 (2014).