Quality control of mitochondrial proteome

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Mitochondria fulfill a plethora of different tasks, which are essential for cell survival. In order to fulfill these functions mitochondria depend on the import of 99% of their protein content. These proteins are synthesized as precursors on cytosolic ribosomes and imported into the target organelle. The translocase of the outer membrane (TOM complex) forms the entry gate for the vast majority of the precursor proteins. Subsequently, dedicated protein machineries sort the precursor proteins into the different mitochondrial subcompartments: the outer and inner membrane, intermembrane space and matrix. Precursor proteins are imported in an unfolded state to pass the TOM channel. Prematurely folded or misfolded precursor proteins can arrest during translocation and cause clogging of the protein translocon. Impaired protein translocation via the TOM complex leads to massive proteotoxic stress. Therefore, the cell harbors molecular mechanisms that extract precursor proteins from the TOM channel and delivers them for proteasomal degradation. Thus, specific quality control factors monitor protein translocases to ensure proper protein import into mitochondria.