

A decline in mitochondrial activity has been associated with aging and is a hallmark of many neurological diseases. Surveillance mechanisms acting at the molecular, organellar, and cellular level monitor mitochondrial integrity and ensure the maintenance of mitochondrial proteostasis. Here we will review the central role of mitochondrial chaperones and proteases, the cytosolic ubiquitin-proteasome system, and the mitochondrial unfolded response in this interconnected quality control network, highlighting the dual function of some proteases in protein quality control within the organelle and for the regulation of mitochondrial fusion and mitophagy ¹⁾.

¹⁾

Baker MJ, Tatsuta T, Langer T. Quality control of mitochondrial proteostasis. Cold Spring Harb Perspect Biol. 2011 Jul 1;3(7). pii: a007559. doi: 10.1101/cshperspect.a007559. Review. PubMed PMID: 21628427; PubMed Central PMCID: PMC3119916.

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