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Sommario/riassunto	Mitochondria are the powerhouses of cells; however, mitochondrial dysfunction causes energy depletion and cell death in a variety of diseases. Altered oxidative phosphorylation and ion homeostasis are associated with ROS production resulting from the disassembly of respiratory supercomplexes and the disruption of electron transfer chains. In pathological conditions, the dysregulation of mitochondrial homeostasis promotes Ca2+ overload in the matrix and ROS accumulation, which induces the mitochondrial permeability transition pore formation responsible for mitochondrial morphological changes linked to membrane dynamics, and ultimately, cell death. Finally, studies on the impaired mitochondrial bioenergetics in pathology could provide molecular tools to counteract diseases associated with mitochondrial dysfunction.