The role of mitochondria in normal and pathological conditions. The possibilities of mitochondrial diseases therapy

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ABSTRACT. Mitochondria are organelles surrounded by a double membrane consisting of outer and inner membranes, an intermembrane space, and a matrix. Mitochondria are involved in a variety of biochemical processes: the synthesis of most of the adenosine triphosphate, the synthesis of fatty acids, the formation of intracellular reactive oxygen species, antioxidant control, oxidative phosphorylation, redox regulation, thermogenesis, the regulation of ionic (calcium in particular) homeostasis, participation in various biosynthetic pathways, and many others, that is why mitochondria are considered the center of cellular metabolism. Mitochondria require constant renewal and replacement of their components for proper functioning and undergo continuous processes of fission, fusion, mitophagy, and transport that determine the morphology, quality, quantity, distribution, and function of mitochondria in cells. Pathology associated with mitochondrial dysfunction can be hereditary or acquired. Primary mitochondrial diseases can arise from mutations in mitochondrial and/or nuclear DNA genes encoding electron transport chain proteins. Secondary mitochondrial dysfunction can accompany many hereditary pathological processes unrelated to oxidative phosphorylation, and can be acquired because of adverse environmental influences that can cause

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oxidative stress. Treatment of mitochondrial diseases is a difficult task, as it is currently practically only symptomatic. Most therapies are aimed either at improving mitochondrial function or at treating the consequences of mitochondrial dysfunction with antioxidants and agents that improve metabolism. Gene therapy and inhibitors of mitochondrial fission are under investigation. The extreme clinical, genetic, and biochemical variability of primary mitochondrial diseases, combined with the small number of patients and the frequent lack of adequate preclinical models, limit the identification of useful clinical outcomes and the development of effective therapies.

KEY WORDS: mitochondria, mitochondria dynamics, metabolism, homeostasis, mitochondrial diseases, treatment.