MITOCHONDRIAL DISORDERS: BIOCHEMICAL BASIS OF DISEASES

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Background. The significance of mitochondria in ensuring cell survival becomes evident through the range of diseases linked to impaired mitochondrial function. For determining the underlying mechanisms and creating specialized treatments, it is essential to comprehend the biochemical basis of mitochondrial disorders. **Objective of the study.** To advance the understanding of the biochemical basis of mitochondrial disorders, which can be used potentially to the development of improved diagnostic methods and therapeutic strategies. **Material and methods.** The research is based on 30 bibliographic sources that were identified using PubMed and NCBI databases, published within the period of 2012-2022. **Results.** Investigations using biochemical methods have shown several important anomalies connected to mitochondrial diseases. Reactive oxygen species accumulation, decreased ATP synthesis, changed mitochondrial membrane potential, and deficits in respiratory chain enzymes are a few of them. Additionally, particular mitochondrial diseases have been linked to abnormalities in the tricarboxylic acid cycle, amino acid metabolism, and fatty acid oxidation. The accumulation of fatty acids can have toxic effects, resulting in the impairment of mitochondrial bioenergetics and disturbances in calcium homeostasis. This, in turn, can induce the opening of permeability transition pores. **Conclusion.** Understanding the molecular basis of mitochondrial disorders is crucial for accurate diagnosis, prognosis, and development of new therapies. Genetic testing, functional assays, and targeted treatments to enhance energy production or reduce oxidative stress offer potential in treating these complex disorders. **Keywords:** mitochondria, mitochondrial disorders, adenosine triphosphate