

Mitochondrial diseases / Disorder of mitochondrial biogenesis

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-----> In the case of a healthy person, the level of energy supply mitochondria is 100% at a young age, gradually decreasing with age, for example around the age of 100 (this is relatively individual, each may be different) will drop to a level at which difficulties may manifest. The most sensitive tissue is brain (damage at about 60% of the "energy service" level), then heart (40%), endocrine organs and kidneys.

In humans with mitochondrial disease, there is a different distribution of mutant mitochondria, which is referred to as "heteroplasmy". This corresponds to different levels of energy supply in different sections of tissue. However, even at a young age, it is less than 100% and gradually decreases due to aging and the potential replication advantage of mutant mitochondria. It depends on when and in which tissue it falls below the threshold characteristic of it, in which the disease manifests itself in damage to the organ.

Thus, mitochondrial diseases oxidative phosphorylation may manifest:

- any symptom,
- in any body,
- at any age.