



Mitochondria

Mitochondria are a membrane-bound organelles found in most eukaryotic cells. These structures are sometimes described as "cellular power sites" because they generate most of the cell's supply of **adenosine triphosphate** (ATP), used as a source of energy. Mitochondria are most plentiful in cells that require significant amounts of energy to function, such as liver and muscle cells.

The mitochondria also involved in other cellular activities like signaling, cellular differentiation, cell senescence and also control of cell cycle and cell growth. Mitochondria also affect human health, like mitochondrial disorder and cardiac dysfunction and they also play important role in the aging process. The term 'mitochondrion' is derived from a Greek word 'mitosis' which means 'thread' and 'chondrion' which means 'granule'.

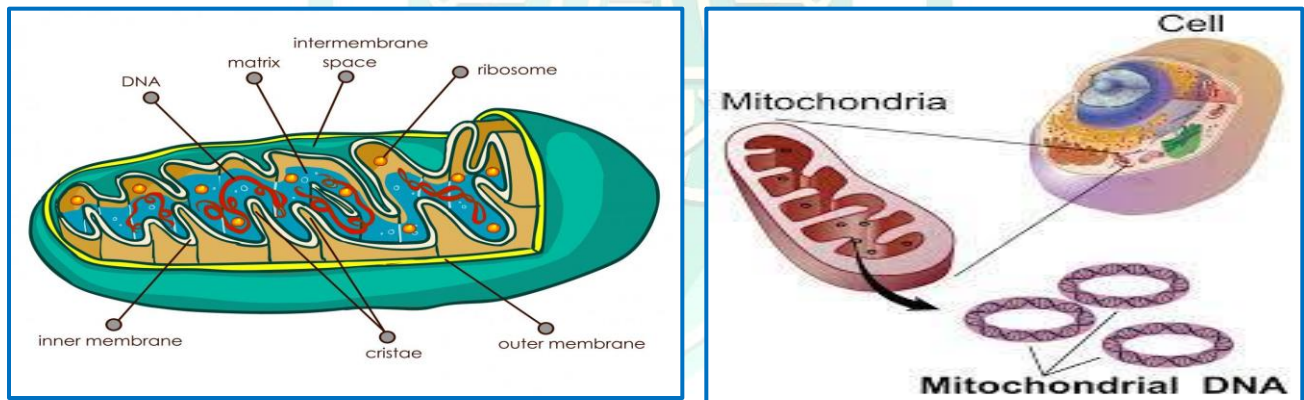
The structure of mitochondria:

Each mitochondrion consists of :

- 1. Outer membrane:** is smooth membrane surrounded that allows entry of molecules and contain enzyme involved in mitochondrial lipid synthesis.
- 2. Intermembrane space:** Because of channels in the outer membrane of the mitochondria, the content of the intermembrane space is similar to that of the content of the cytoplasm.
- 3. Inner membrane:** exhibit numerous folds called **cris**tae which maximize internal surface area of mitochondria and contain most of the respiratory chain enzymes and

ATP synthase which is responsible for cell respiration (**oxidative phosphorylation**) and production of cell ATP. Shape of cristae different according type of cells; in protein secreting cells cristae project into the interior of the organelle like shelf. In steroid secreting cells such as the adrenal cortex or interstitial cells in the testes, the mitochondria cristae are tubular.

4. Mitochondrial matrix: the matrix is the space within the inner membrane; contain enzymes for **Krebs cycle**, **mitochondrial DNA** (circular DNA), **special ribosome**, **tRNase** and **enzymes** for gene expression.



- Mitochondrial DNA is double stranded and has a circular structure very similar to bacterial chromosomes, mitochondrial DNA synthesis and duplication is independent of nuclear DNA replication.
- Mitochondrial ribosome is smaller than cytosolic ribosome.
- tRNases are enzymes that degraded the tRNA.

Content of Mitochondrial Matrix

- ☒ **Matrix granules**, which are thought to represent accumulations of divalent cations, mostly calcium. This suggests that the mitochondria help regulate calcium levels in the cytoplasm.
- ☒ **Various enzymes**, including those of the citric acid cycle
- ☒ **DNA, RNA, and ribosomes**, which are similar to those found in bacteria. This suggests that mitochondria may have evolved from aerobic bacteria that somehow became incorporated into animal cells. In fact, mitochondria divide independently of cell division, further evidence for a once autonomous existence.

Replication of mitochondria

Mitochondria replicate similarly to bacterial cells, when they get large, they undergo fission. This involves furrowing of the inner and then the outer membrane as if someone was pinching the mitochondrion. The two daughter cells of the mitochondria must first replicate the DNA.

Function of mitochondria

1. Mitochondria are primary sites for ATP synthesis (site of Krebs cycle) from organic material so that known as powerhouse of the cell.
2. Cell respiration.
3. Maintain body heat because some energy dissipated as heat.
4. They have key role in apoptosis programmed cell death.
5. Some mitochondrial functions are performed only in specific types of cells, e.g. mitochondria in liver cells contain enzymes that allow them to detoxify ammonia, a waste product of protein metabolism

Mitochondrial DNA

Mitochondrial DNA or mtDNA or mDNA is the DNA in the mitochondria, rest of the DNA present in the eukaryotic cells is in the nucleus, in plants DNA is also found in chloroplasts.

The mitochondria have a small amount of DNA of their own. Human mitochondrial DNA spans about 16,500 DNA base pairs, it represents a small fraction of the total DNA in cells. The mtDNA contains 37 genes. All these genes are essential for normal function of the mitochondria.

These DNA help the mitochondria divide independently from the cell. mtDNA is maternally inherited. The fact that mt DNA is maternally inherited enables to trace the maternal lineage far back in time. The mt DNA in most multicellular organisms is circular, covalently closed, double-stranded DNA. mtDNA is susceptible to free oxygen radicals. Mutations in the mitochondrial DNA leads to a number of illness like exercise intolerance.

Mitochondrial Disease

Disease of mitochondria results due to the failure of mitochondria. Dysfunction in the mitochondria fails to produce energy that is needed for the sustainment of life and growth of an organism. Injury in the cell or even cell death results in the production of less energy. If the process happens throughout the body, the whole system begins to fail. The disease primarily affects young. The mitochondrial disease causes most of the damage to the cells of brain, heart, liver, muscles, kidney, respiratory and the endocrine systems.