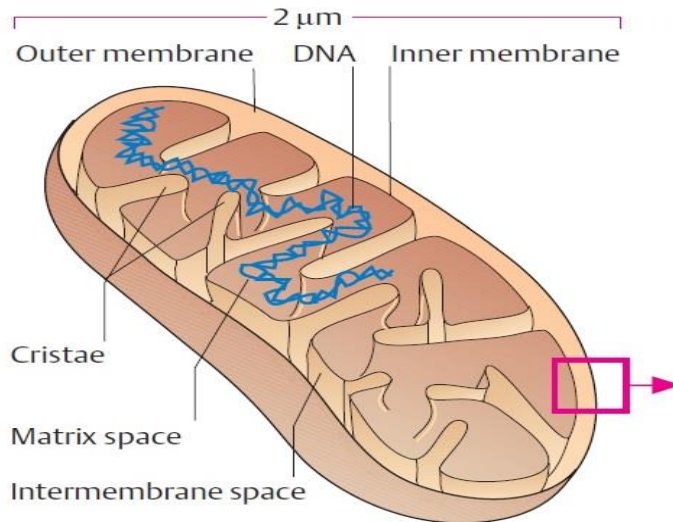


## Mitochondria – Structure and Functions

Mitochondria are bacteria-sized organelles (about  $1 \times 2 \mu\text{m}$  in size), which are found in large numbers in almost all eukaryotic cells. Typically, there are about 2000 mitochondria per cell, representing around 25% of the cell volume.



### Structure of Mitochondria

- Mitochondria are enclosed by two membranes—a smooth outer membrane and a markedly folded or tubular inner mitochondrial membrane, which has a large surface and encloses the matrix space.
- The folds of the inner membrane are known as cristae, and tube-like protrusions are called tubules.
- The intermembrane space is located between the inner and the outer membranes.
- The number and shape of the mitochondria, as well as the numbers of cristae they have, can differ widely from cell type to cell type.
- Tissues with intensive oxidative metabolism— e. g., heart muscle—have mitochondria with particularly large numbers of cristae.
- Even within one type of tissue, the shape of the mitochondria can vary depending on their functional status.
- Mitochondria are mobile, plastic organelles.
- Mitochondria probably developed during an early phase of evolution from aerobic bacteria that entered into symbiosis with primeval anaerobic eukaryotes.
- This endo-symbiont theory is supported by many findings. For example, mitochondria have a ring-shaped DNA (four molecules per mitochondrion) and have their own ribosomes. The mitochondrial genome became smaller and smaller during the course of evolution.
- In humans, it still contains 16,569 base pairs, which code for two rRNAs, 22 tRNAs, and 13 proteins.

- Only these 13 proteins (mostly subunits of respiratory chain complexes) are produced in the mitochondrion.
- The mitochondrial envelope consisting of two membranes also supports the endosymbiont theory.
- The inner membrane, derived from the former symbiont, has a structure reminiscent of prokaryotes.
- It contains the unusual lipid cardiolipin, but hardly any cholesterol.
- Both mitochondrial membranes are very rich in proteins.
- Porins in the outer membrane allow small molecules to be exchanged between the cytoplasm and the intermembrane space.
- The inner mitochondrial membrane is completely impermeable even to small molecules (with the exception of O<sub>2</sub>, CO<sub>2</sub>, and H<sub>2</sub>O).
- Numerous transporters in the inner membrane ensure the import and export of important metabolites.
- The inner membrane also transports respiratory chain complexes, ATP synthase, and other enzymes.
- The matrix is also rich in enzymes.

## Mitochondria

Mitochondria are well-defined cytoplasmic organelles of the cell which take part in a variety of cellular metabolic functions. Survival of the cells requires energy to perform different functions. The mitochondria are important as the fact that these organelles supply all the necessary biological energy of the cell, and they obtain this energy by oxidizing the substrates of the Krebs cycle. Energy of the cell is got from the enzymatic oxidation of chemical compounds in the mitochondria. Hence, the mitochondria are referred to as the 'power houses' of the cell. Almost all the eukaryotic cell have mitochondria, though they are lost in the later stages of development of cell like in the red blood cells or in elements of phloem sieve tube.

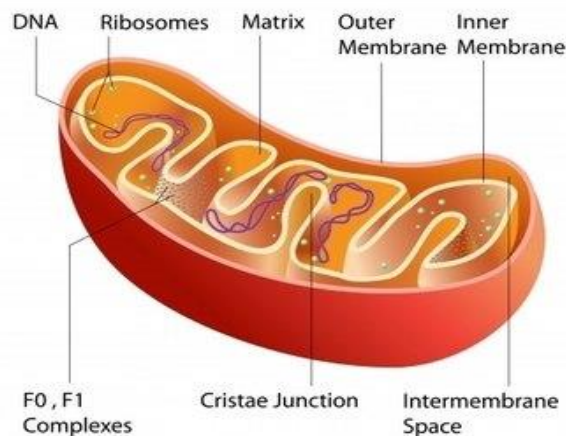
In 1890, mitochondria was first described by Richard Altmann and he called them as bioblasts. Benda in the year 1897 coined the term mitochondrion. In the 1920s, a biochemist Warburg found that oxidative reactions takes place in most tissues in small parts of the cell.

### Mitochondria Definition

Mitochondria is a membrane bound cellular structure and is found in most of the eukaryotic cells. The mitochondria ranges from 0.5 to 1.0 micrometer in diameter. The mitochondria are sometimes described as power plants of the cells. These organelles generate most of the energy of the cell in the form of adenosine triphosphate (ATP) and it is used as a source of chemical energy. 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'.

### Structure of Mitochondria

Mitochondria are rod shaped structure found in both animal and plant cells. It is a double membrane bound organelle. It has the outer membrane and the inner membrane. The membranes are made up of phospholipids and proteins.



The components of mitochondria are as follows:

### **Outer membrane**

- It is smooth and is composed of equal amounts of phospholipids and proteins.
- It has a large number of special proteins known as the porins.
- The porins are integral membrane proteins and they allow the movement of molecules that are of 5000 daltons or less in weight to pass through it.
- The outer membrane is freely permeable to nutrient molecules, ions, energy molecules like the ATP and ADP molecules.

### **Inner membrane**

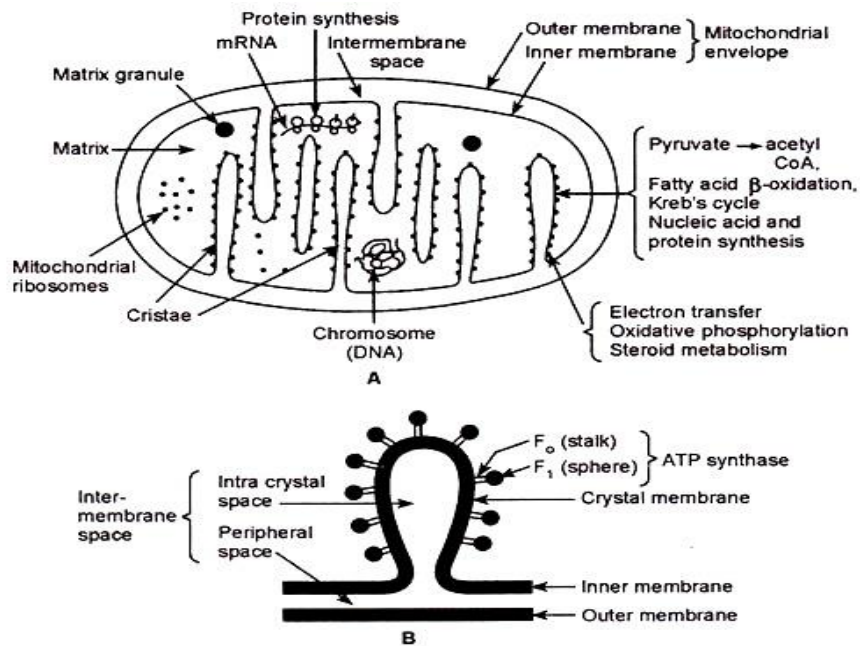
- The inner membrane of mitochondria is more complex in structure.
- It is folded into a number of folds many times and is known as the cristae.
- This folding help to increase the surface area inside the organelle.
- The cristae and the proteins of the inner membrane aids in the production of ATP molecules.
- Various chemical reactions takes place in the inner membrane of the mitochondria.
- Unlike the outer membrane, the inner membrane is strictly permeable, it is permeable only to oxygen, ATP and it also helps in regulating transfer of metabolites across the membrane.

### **Intermembrane space**

- It is the space between the outer and inner membrane of the mitochondria, it has the same composition as that of the cell's cytoplasm.
- There is a difference in the protein content in the intermembrane space.

### **Matrix**

- The matrix of the mitochondria is a complex mixture of proteins and enzymes. These enzymes are important for the synthesis of ATP molecules, mitochondrial ribosomes, tRNAs and mitochondrial DNA.



**Fig. 2.21.** Diagrammatic representation of mitochondrion ultrastructure. **A.** Mitochondrial cross section showing various parts. **B.** Detailed structure of a crista.

## Function of Mitochondria

Functions of mitochondria depends on the cell type in which they are present.

- The most important function of the mitochondria is to produce energy. The simpler molecules of nutrition are sent to the mitochondria to be processed and to produce charged molecules. These charged molecules combine with oxygen and produce ATP molecules. This process is known as oxidative phosphorylation.
- Mitochondria help the cells to maintain proper concentration of calcium ions within the compartments of the cell.
- The mitochondria also help in building certain parts of blood and hormones like testosterone and estrogen.
- The liver cells mitochondria have enzymes that detoxify ammonia.
- The mitochondria also play important role in the process of apoptosis or programmed cell death. Abnormal death of cells due to the dysfunction of mitochondria can affect the function of organ.

## 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 mtDNA 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.

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