CELL BIOLOGY

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Lecture 8: Mitochondria 22/11/2021

Mitochondria are membrane-enclosed organelles distributed through the cytosol of most eukaryotic cells. Their number within the cell ranges from a few hundred to, in very active cells, thousands. Their main function is the conversion of the potential energy of food molecules into ATP.

• The individual mitochondrion is a bilayer structure and contains four compartments:

***Inner membrane**

The **inner membrane** contains a variety of enzymes. It contains ATP synthase which generates ATP in the matrix, and transport proteins that regulate the movement of metabolites into and out of the matrix.

The inner membrane is arranged into **cristae** in order to increase the surface area available for energy production via **oxidative phosphorylation**.

***Outer membrane**

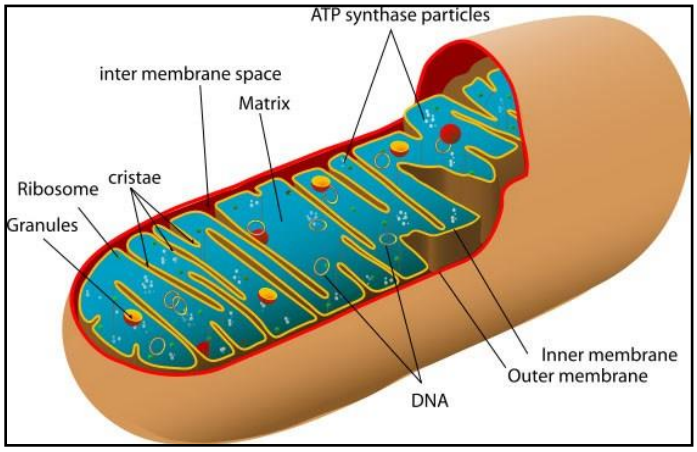
The outer membrane covers the surface of the **mitochondrion** and has a large number of special proteins known as porins. It is freely permeable to ions, nutrient molecules, energy molecules like the ADP and ATP molecules.

***Matrix**

The space within the inner membrane of the mitochondrion is known as the **matrix**, which contains the enzymes of the Krebs (TCA) and fatty acid cycles, alongside DNA, RNA, ribosomes and calcium granules.

***Intermembrane space**

Space between inner and outer membranes



Why 2 membranes? increase surface area for membrane-bound enzymes that synthesize ATP

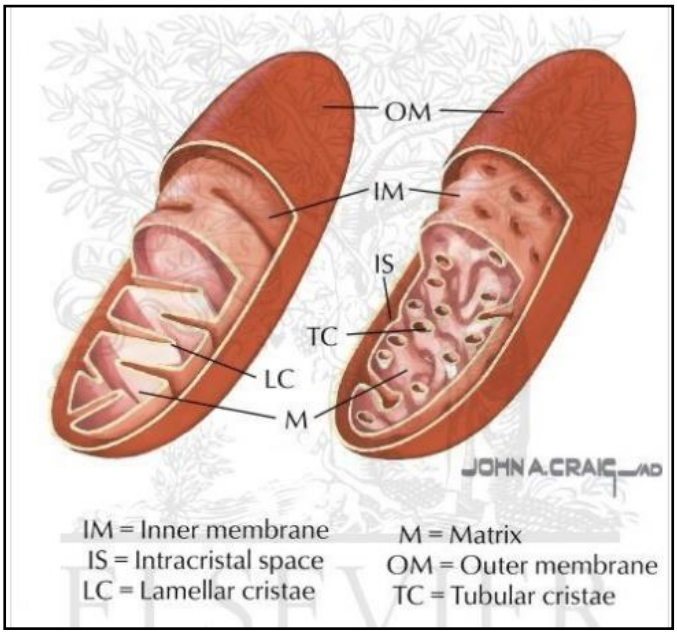
Isolated mitochondria have two morphologic forms according to activity:

1-O<u>rthodox form (inactive form)</u> mitochondria inactive in oxidative phosphorylation **(expanded, less dense matrix and more compact cristae)**

2-<u>Condensed mitochondrial form (active form)</u> . In this form the volume of the outer chamber is increased to approximately 50% of the organelle, and the inner chamber is reduced in volume. **(contracted, very dense matrix and wide cristae)**

Types of mitochondria according to cristae:

1) Lamellar mitochondria: arranged in shelves e.g. parietal gastric cell
2) Tubular mitochondria: the cristae appear as finger like, e.g. steroid secreting cells



The number and size of mitochondria are, in general, correlated with the level of oxidative phosphorylation.

More mitochondria are found in cells that:
-have motile machinery
-sequester low pH substances
-pump large amounts of ions

Cells known to have high number of mitochondria

- **Hepatocyte** may each contain about 1,000 to 1,500 mitochondria
- **Cardiac muscles**
- **Skeletal Muscle (**striated muscle, which contain myofilaments that slide on one another to effect contraction, mitochondria are present close to the myofilaments).
- **Parietal cell of the stomach**
- **Ciliated cells: ATP is needed to move the microtubules in the cilia, Cilia helps move mucus along in the lumen of the trachea and the ovum in the oviduct**
- **Presynaptic terminal of neuron**
- **Tail of sperm**
- **Cells of proximal convoluted tubule**

Mature erythrocytes, totally dependent for energy on glycolysis, contain none.

Small lymphocytes have a few number.

Once inside the cell, glucose is broken down to make ATP in two pathways:-

1- Anaerobic metabolism: pathway requires no oxygen called glycolysis and it occur in the cytoplasm, outside of mitochondria. During glycolysis, glucose is broken down into pyruvate.

2- Aerobic respiration takes place in the mitochondria in the presence of oxygen. Pyruvic acid is converted into carbon dioxide, and water thus releasing a lot of energy.

That is why mitochondria and oxygen are so important. We need to continuo the breakdown with the Krebs cycle inside the mitochondria in order to enough ATP to run all the cell function.

Cellular respiration:

Three Main Stages

- **Glycolysis (2 ATP)**
- **Kreb's Cycle (2 ATP)**
- **Electron Transport Chain (32 ATP)**

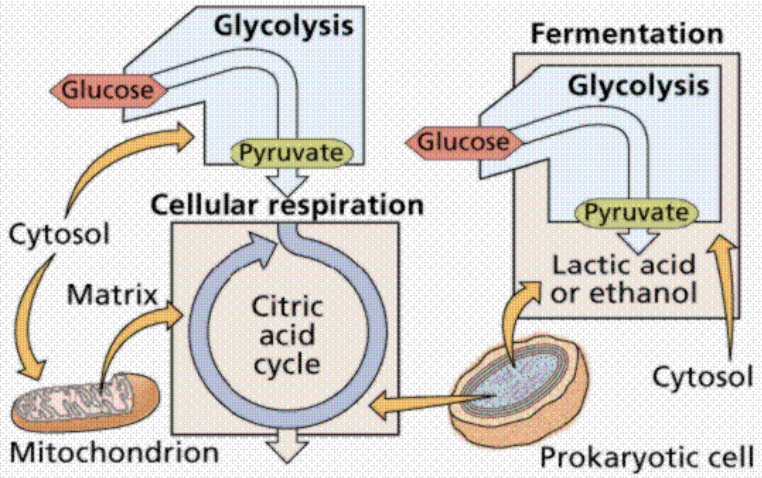


Figure : Anaerobic and aerobic metabolism

Mitochondrial DNA (mtDNA)

Inside the mitochondrion is a certain type of DNA. That's different in a way from the DNA that's in the nucleus.

-This DNA is small and circular.

-It has only 16,500 or so base pairs in it.

-It encodes different proteins that are specific for the mitochondrial function

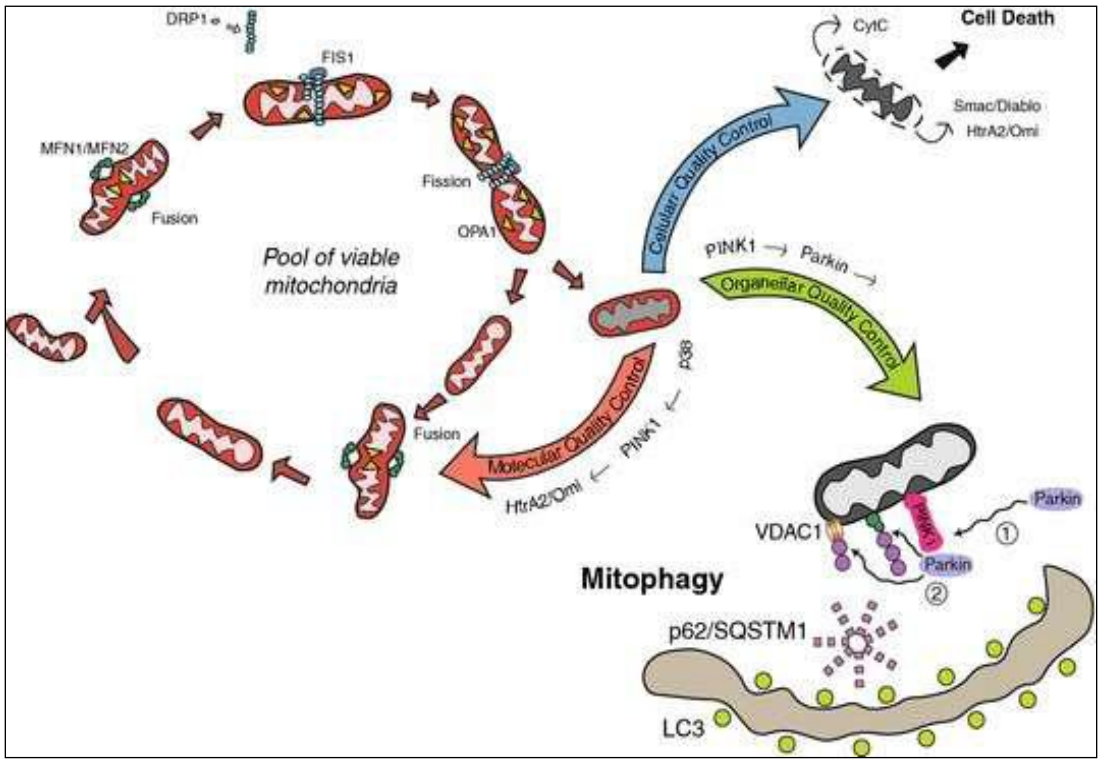
-Some of the enzymes in those pathways, and some of the proteins that are needed to function in those pathways, are produced by the mitochondrial DNA.

The mitochondrial DNA is critically important for many of the pathways that produce energy within the mitochondria. And if there's a defect in some of those mitochondrial DNA bases, that is to say a mutation, it causes mitochondrial disease, which will involve the inability to produce sufficient energy in things like the muscle and the brain, and the kidney.

The parts of the body, such as the heart, brain, muscles and lungs, requiring the greatest amounts of energy are the most affected during mitochondrial disease.

Mitochondria life cycle: fusion, fission and autophagy

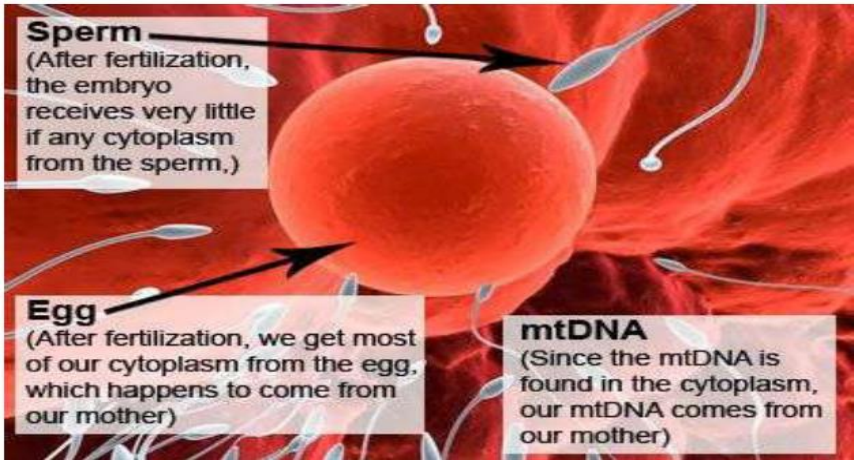
Mitochondria cant be synthesized de novo, so new mitochondria must arise from existing mitochondria.
At any point of time, mitochondria are in a dynamic flux between fission and fusion.



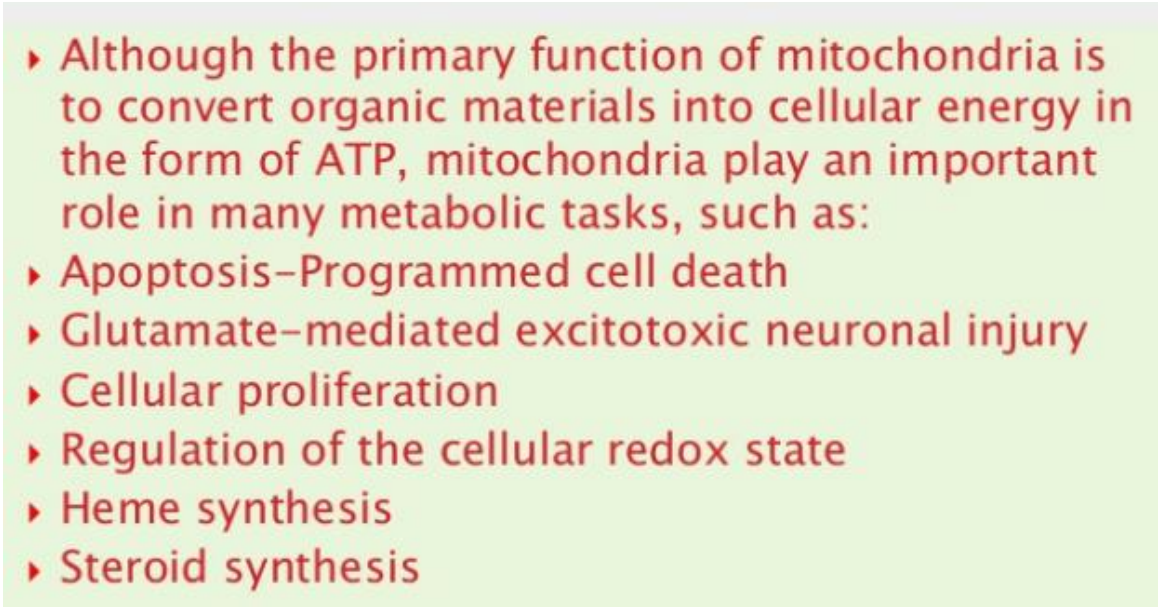
Mitophagy is the selective degradation of mitochondria by autophagy. It often occurs to defective mitochondria following damage or stress.

Maternal inheritance: In sexual reproduction, mitochondria are normally inherited exclusively from the mother; the mitochondria in mammalian sperm are usually destroyed by the egg cell after fertilization.

MATERNAL INHERITANCE



Additional functions



Some mitochondrial functions are performed only in specific types of cells. For example, mitochondria in liver cells contain enzymes that allow them to detoxify ammonia, a waste product of protein metabolism. A mutation in the genes regulating any of these functions can result in mitochondrial diseases.