



## Mitochondria

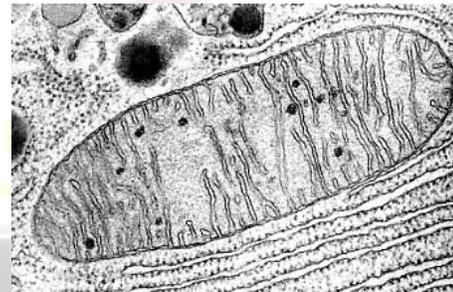
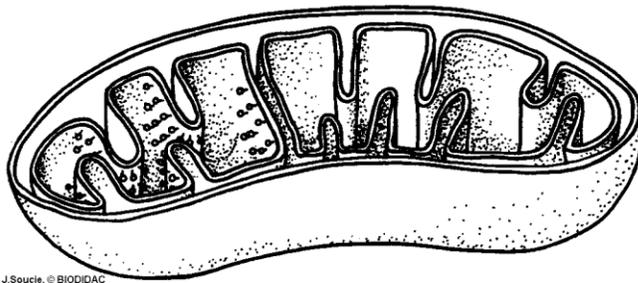
By

Dr. Barrister Kumar Gupta

B.Sc.I, SEM II, Paper1- Unit-II

Cell Biology & Genetics

Mitochondria are small<sup>1</sup>, rod-shaped, semiautonomous intracellular organelles responsible for energy production and cellular respiration in all eukaryotic cells except mature RBCs of human cells. The mitochondria of a cell are collectively designated as **chondriome**.



### Historical<sup>2</sup>:

Mitochondria were first observed, and called “sarcosomes”, by Kolliker (1857). Later these were renamed as “bioblasts” by Altmann (1894) and “mitochondria” by C. Benda (1897).

### Ultrastructure:

All mitochondria consist of two membranes and two compartments. An outer membrane surrounds the mitochondrion; within this membrane and separated from it is an inner membrane that projects into the mitochondrial cavity as complex infolding called mitochondrial crests. This inner membrane divides the mitochondrion into two compartments: (1) the outer peri-mitochondrial chamber contained between the outer and

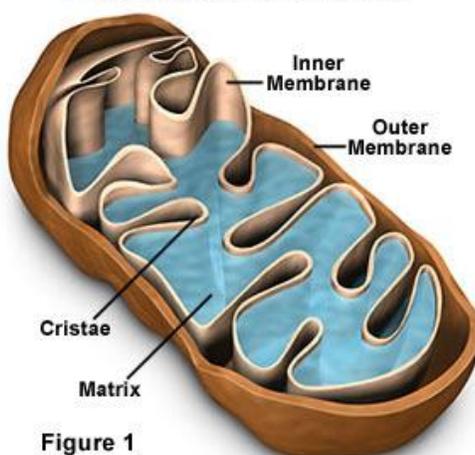
<sup>1</sup> They are large enough to be seen under a light microscopes<sup>1</sup>; visibility is increased by the vital stain, **Janus green**.

<sup>2</sup> Kingsbury (1912) first suggested that mitochondria were the sites of cellular respiration. Palade and Sjostrand (1940-50) worked out the fine structure of the mitochondria under the electron microscope. Claude (1944) separated the mitochondrial fraction from other cell components by ultracentrifugation.



inner membranes and in the core of crests and (2) the inner chamber inside the inner membrane which is filled with relatively dense proteinaceous material, called mitochondrial matrix. Within the mitochondrial matrix are small ribosomes, a circular mitochondrial DNA and enzymes of the Krebs cycle.

Mitochondria Inner Structure



All the enzymes of the electron transport system are found embedded within the inner membrane. In addition, the M face (facing matrix) of the inner membrane in the crests has regularly spaced units of stalked particles called “**elementary particles**” or “**F1 particles**”<sup>3</sup>. These particles correspond to special ATPase involved in the coupling of oxidation and phosphorylation.

**Chemical composition:**

Both membranes have trilamellar structure; however, the outer membrane has a much higher lipid-protein ratio than the inner membrane.

**Mitochondrial genome:**

Mitochondrial DNA (mtDNA) is a *circular, double stranded, histone-free and membrane bound molecule*, reflecting their bacterial origin. A single mitochondrion may contain 2 to 6 DNA molecules<sup>4</sup>. The mtDNA codes for the mitochondrial rRNAs, tRNAs, and some of the essential mitochondrial proteins. Most of the mitochondrial proteins are, however, encoded by the nuclear genes, synthesized in the cytosol, and transferred into

<sup>3</sup> Also called “oxysomes” or subunits of Fernandez-Moran.

<sup>4</sup> Depending on its size; i.e. the larger the mitochondrion, the more DNA molecules present.



the various components of mitochondrion through the contact zones<sup>5</sup> by a post transcriptional mechanism.

Because in mammals and most multicellular organisms, sperm contributes little (if any) cytoplasm to the zygote, virtually all the mitochondria in the embryo are derived from those in the egg; thus *inheritance of mitochondria is maternal*.

Since the mitochondria (and chloroplasts) resemble very closely prokaryotic cells with respect to size, distribution of respiratory enzymes, and the striking similarity of their DNA and RNA components, it has been speculated that perhaps (both chloroplasts and) mitochondria originated from prokaryotic endosymbionts that, over a long evolutionary period, were gradually integrated into their host (symbiont hypothesis).

## Endosymbiosis

Mitochondria formed as a result of an endosymbiotic event around 2 billion years ago.

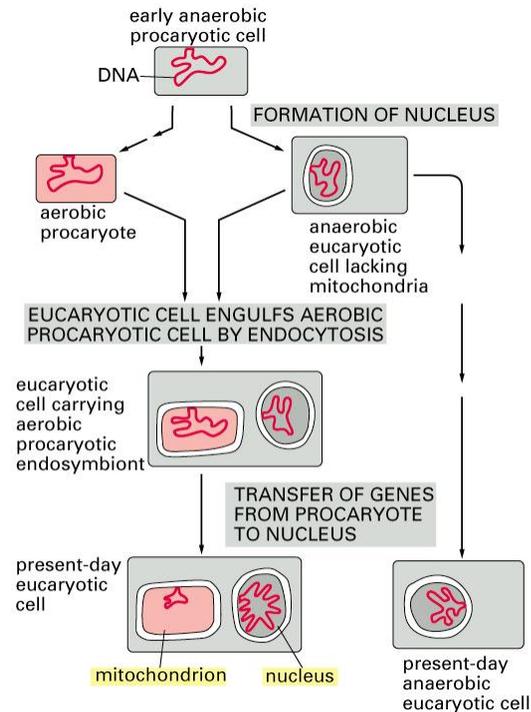


Figure 14-56. Molecular Biology of the Cell, 4th Edition.

### Function:

The mitochondria function as *energy-transducing organelles* into which the major degradation products of cell metabolism penetrate and are converted into chemical energy (ATP) to be used in various activities of the cell. They are often referred to as the

<sup>5</sup> Where two membranes of the mitochondria associate.



'powerhouse' of the cell since they produce 95% of ATP molecules in animal cells<sup>6</sup>. The production of energy (i.e., cellular respiration) is based on three coordinated steps- 1) the Krebs cycle, 2) the electron transport system, and 3) the phosphorylating system.

**1. The Krebs cycle:** The Krebs cycle, which is a cyclic system of enzymatic reactions for the oxidation of acetyl residues in the mitochondrial matrix, produces CO<sub>2</sub> by decarboxylation and removes electrons from the metabolites.

**2. The respiratory chain or electron transport system:** It captures the pairs of electrons and transfers them through a series of electron carriers to activated oxygen. It may be divided into four multi-molecular complexes<sup>7</sup>:

1) **Complex 1 or NADH-Q-reductase:** It catalyzes the reduction of CoQ by electrons removed from NADH.

2) **Complex 2 or Succinate-Q-reductase:** It catalyzes the reduction of CoQ by electrons removed from succinate.

3) **Complex 3 or QH<sub>2</sub>-cytochrome-c reductase:** It catalyzes the reduction of cytochrome-c by electrons removed from CoQ.

4) **Complex 4 or cytochrome c oxidase:** It catalyzes the oxidation of reduced cytochrome c by molecular oxygen.

**3. Phosphorylating system:** The phosphorylating system is represented by the F<sub>1</sub>-ATPase, a multiprotein complex with 3 main parts: 1) the F<sub>1</sub> particle or soluble ATPase, 2) the proton translocating portion (F<sub>o</sub>) which is a complex of very hydrophobic proteins and 3) a protein stalk that connects the two and contains the coupling factors. It is tightly coupled in the respiratory chain, which at three points give rise to ATP molecules.

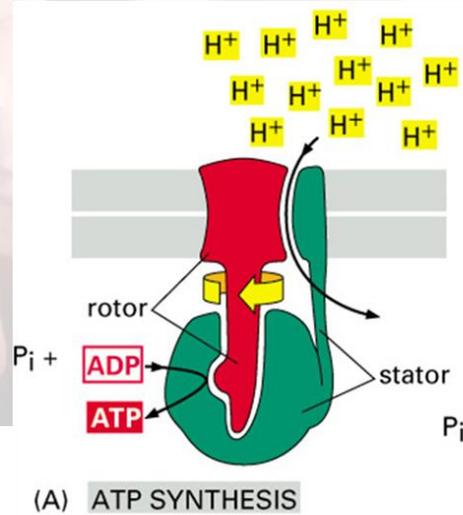


Figure 14–19. Molecular Biology of the Cell,

<sup>6</sup> 5% ATP is produced during anaerobic respiration outside the mitochondria.

<sup>7</sup> In which lipids are essential for activity.



The nature of link between the respiratory chain and the phosphorylating system (or  $F_1$ -ATPase) is unknown, but the chemiosmotic theory of P. Mitchell which proposes an electrochemical link between respiration and phosphorylation is the most favoured one.

**The chemiosmotic theory:** According to this theory, the inner membrane of the mitochondrion acts as a transducer converting the energy provided by an electrochemical gradient into the chemical energy of ATP. It states that a proton gradient is established as electrons flow from NADH to  $O_2$  along the respiratory chain and the disestablishment of this gradient (due the return of these electrons from C side back to M side of the membrane through the proton channel of  $F_1$ ATPase) results in the synthesis of ATP. For each NADH that is oxidized three molecules of ATP are generated.

Images:

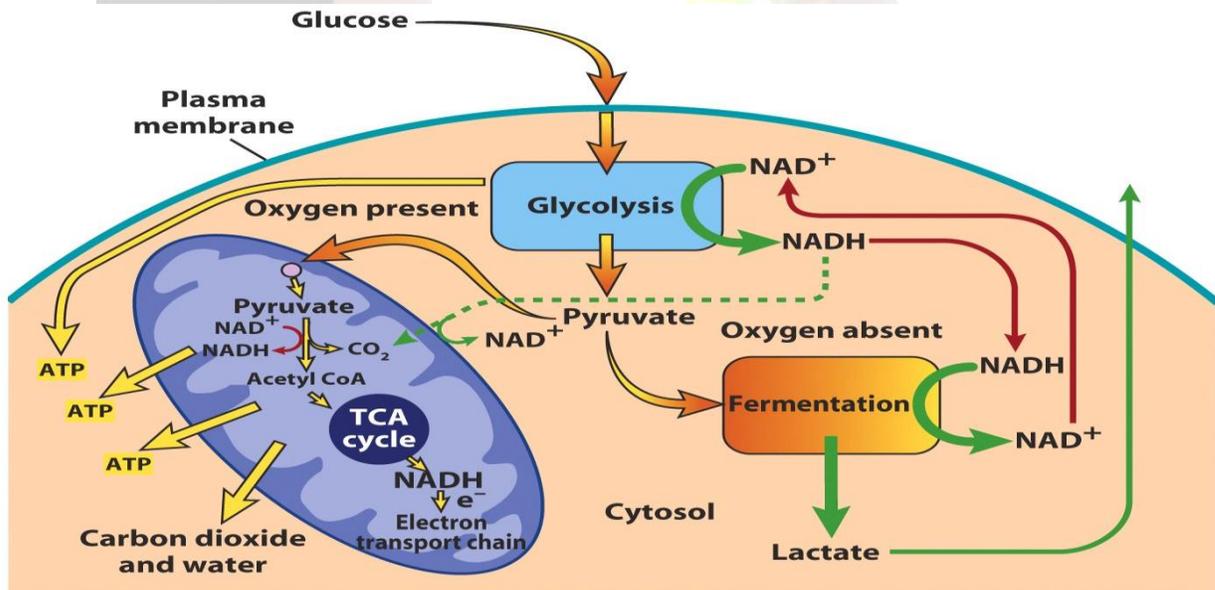
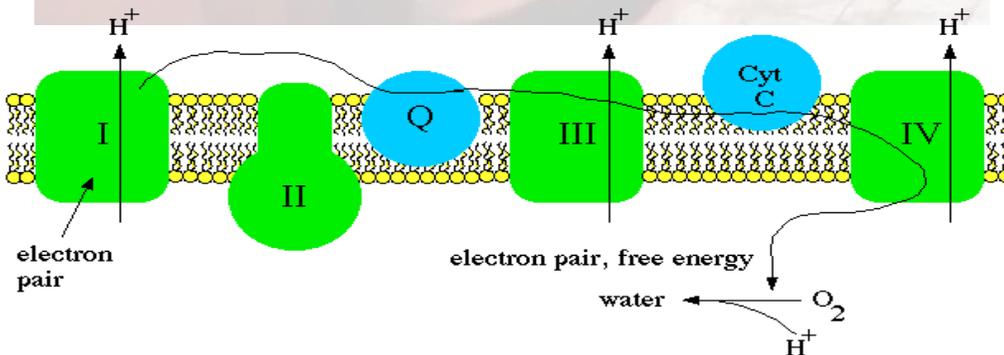
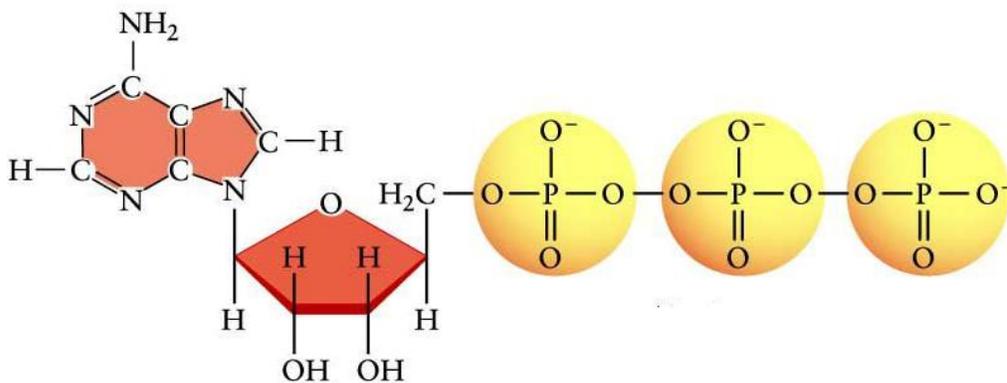
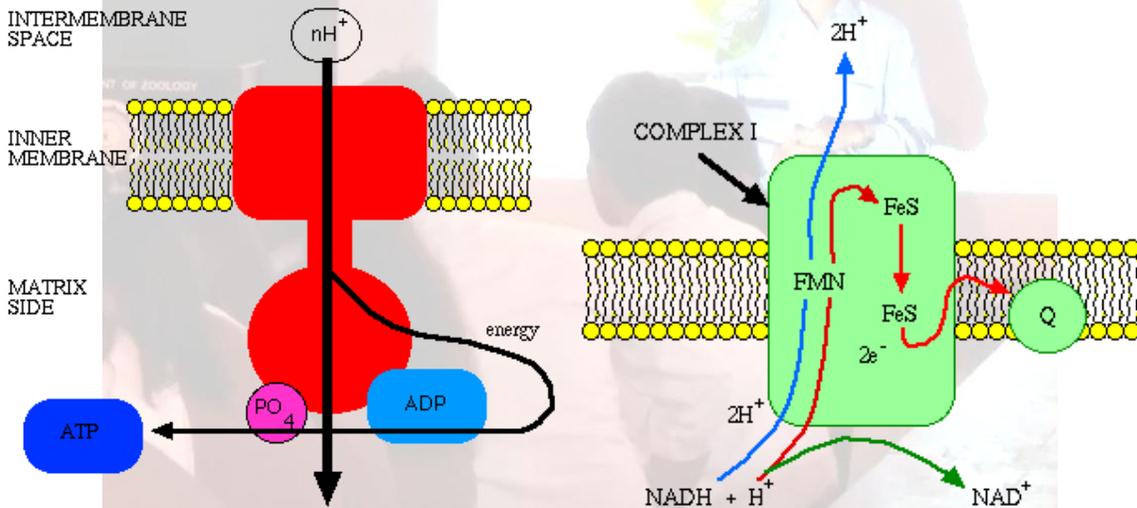
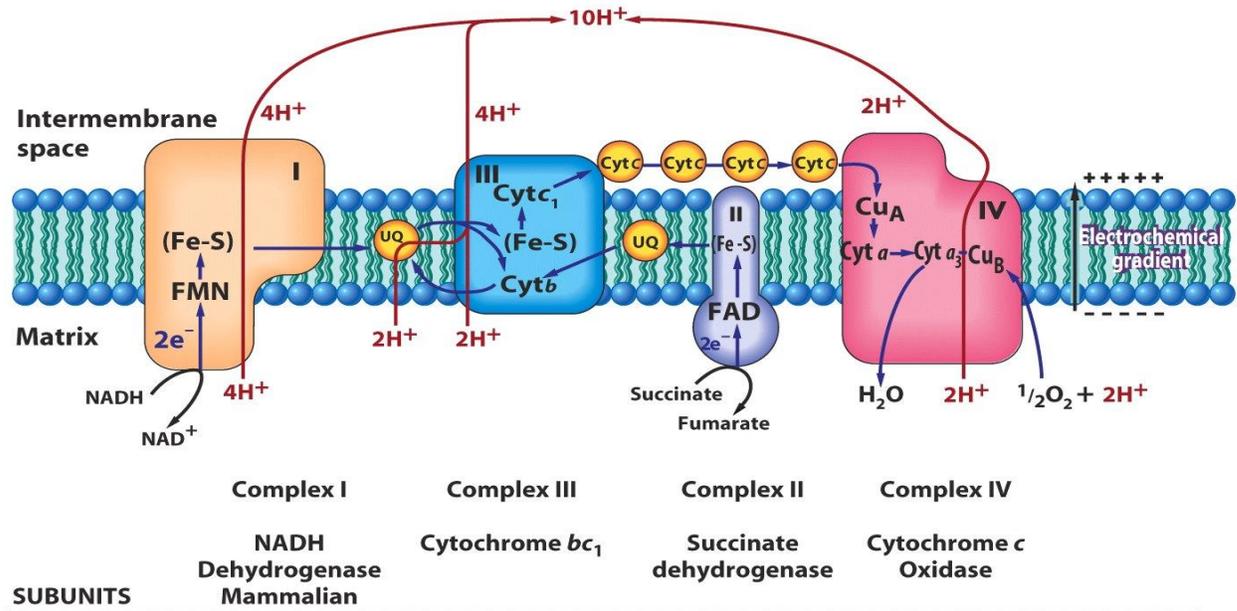


Figure 5-5 Cell and Molecular Biology, 4/e (© 2005 John Wiley & Sons)





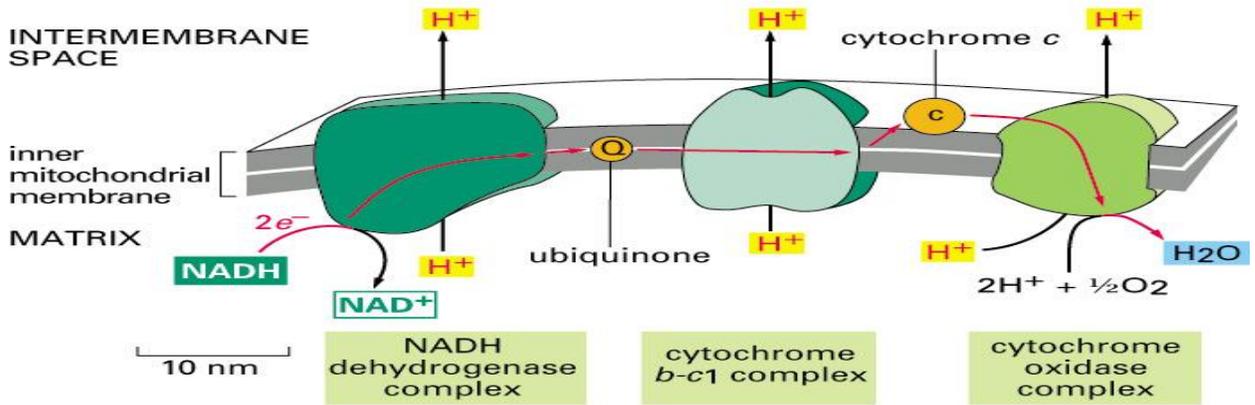


Figure 14–26. Molecular Biology of the Cell, 4th Edition.

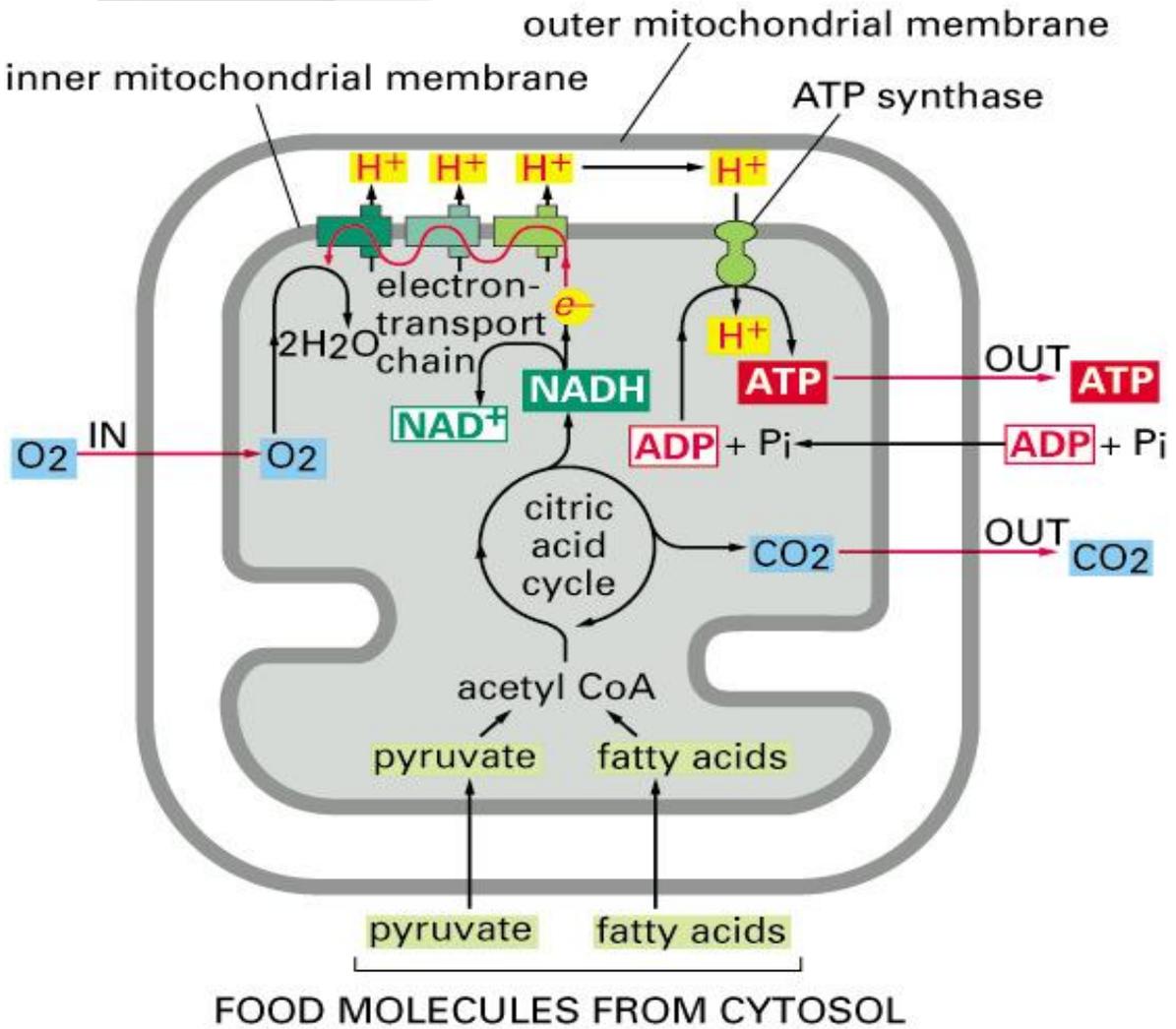
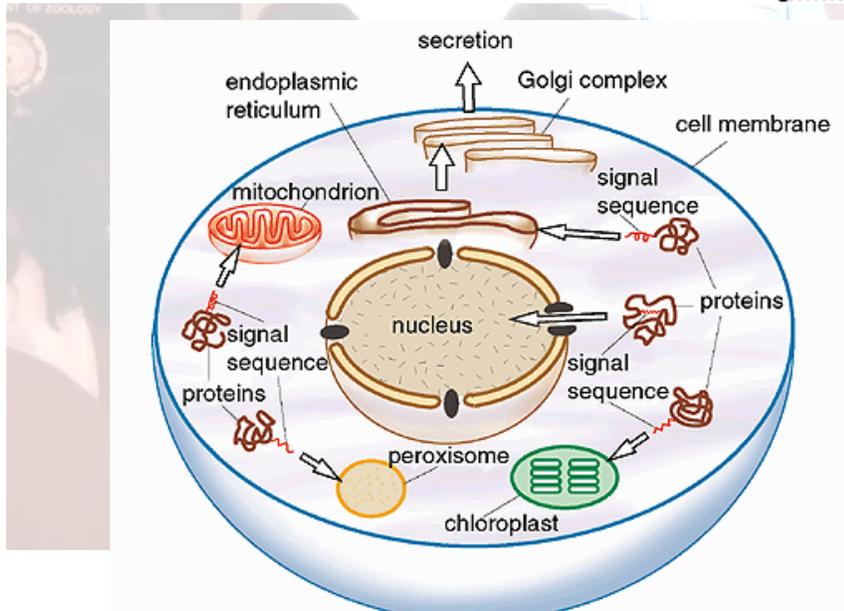
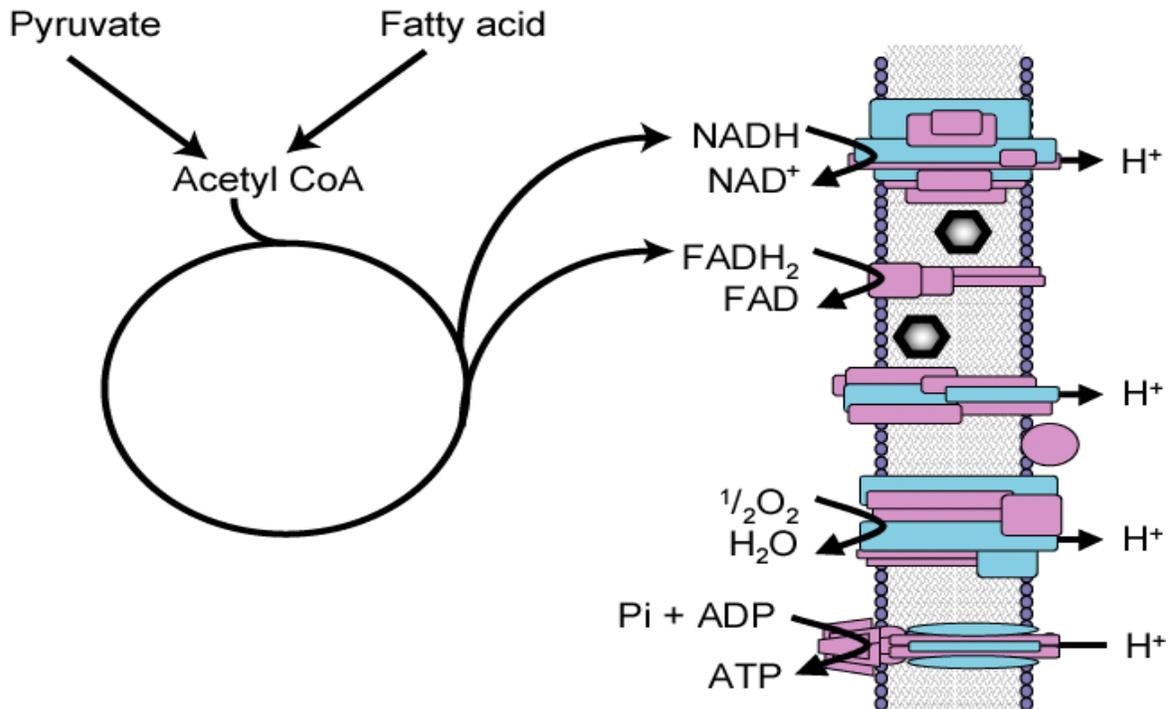


Figure 14–10. Molecular Biology of the Cell, 4th Edition.



### Textbooks

- Alberts B, Johnson A, Lewis J, et al. Molecular Biology of the Cell. 4<sup>th</sup> edition. New York: Garland Science; 2002.
- Cooper GM. The Cell: A Molecular Approach. 2nd edition. Sunderland (MA): Sinauer Associates; 2000. Mitochondria.