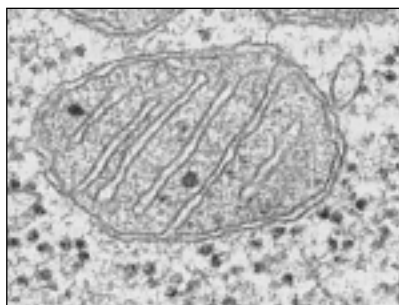


Mitochondria

Mitochondria are free floating organelles within the cytosol (cytoplasm) which have been variously described as 'the powerhouses of the cell' or 'the electric company of the cell'. They are in the business of producing energy for cellular respiration.

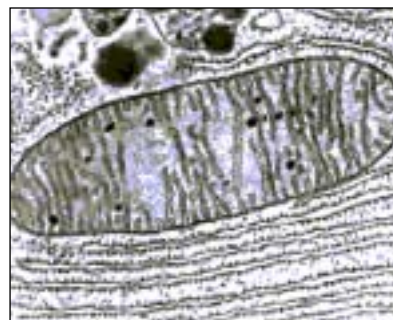


Floating inside the cytoplasm are a series of fairly large organelles called Mitochondria. These organelles, the size of some bacteria, serve as the cell's respiration centres, the place where energy for the cell is produced. Since the Mitochondria serves as a centre for energy production, there are varying numbers of mitochondria in different cells. Muscles have many mitochondria due to the amount of energy they need, but skin cells have very few.

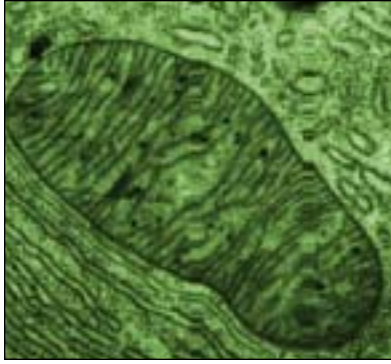
The mitochondria (singular: mitochondrion) have two membranes. The outer membrane protects the organelle, and contains specialized transport proteins such as porin which allows free passage for various molecules into the intramembranous space of the mitochondria (the space between the inner and outer membranes). The inner membrane is folded into a series of cristae or long folds and is highly impermeable to small ions due to having a very high content of a phospholipid called cardiolipin.

The folds partially fill the inner cavity or matrix and greatly increase the surface area of the inner membrane allowing more area for energy production. This matrix serves as a place for the chemical reactions involved in energy production and contains respiratory chain enzymes as well as ATP (Adenosine Tri-Phosphate) synthetase the precursor to ATP or Adenosine triphosphate.

The production of ATP takes place in the mitochondria by a process known as oxidative phosphorylation.



During the complete metabolism of 1 molecule of glucose in the mitochondria 38 molecules of ATP are produced each of which stores energy in phosphate bonds. Most of these and thus most of the body's store of energy are produced during a very important biological process known as the Krebs Cycle. The energy stored in the ATP is released by converting it to ADP or Adenosine diphosphate. The conversion of 1 mole (equivalent to the molecular weight in grams) of ATP to ADP releases 7kcal of energy.



Electron Micrograph of mitochondria
(x80,000 approx.)

In the human being all mitochondria derive from mitochondria present in the maternal ovum. There is no contribution from the father's sperm. Sometimes the mitochondria divide abnormally in the developing foetus leading to various conditions known collectively as mitochondrial cytopathy syndromes.

The most common patterns of disease shown are :

- Muscle weakness particularly affecting the extraocular muscles which control eye movement.
- Degenerative disease of the central nervous system.
- Metabolic disorders usually manifesting as abnormally high levels of lactic acid.

Such disease may present at any time from childhood to adulthood.

The severity of the disease depends on the number of mitochondria affected. Individuals may suffer from a condition known as mosaicism where some of the mitochondria are normal and some are abnormal. The higher the proportion of abnormal mitochondria the more severe the symptoms of the disease.

Diagnosis can be aided by examining the muscle tissue cells via electron microscopy for abnormal mitochondria.

Much scientific debate has ensued over whether or not the mitochondria are really part of your cell, or just a helpful parasite. The fact that they contain their own DNA and ribosomes, and that they divide when they want to divide, and not when the rest of the cell divides, suggests that they are in fact a separate entity inside the cell. One widely accepted theory states that Mitochondria are in fact evolved from prokaryotic cells similar to bacteria, and they have developed a symbiotic relationship with the eukaryotic cell. Receiving protection as a reward for giving energy! So the mitochondria may be the cell's very own X-files!