Mitochondria are vital organelles within cells, often called the "powerhouses" because they generate most of the cell's energy in the form of ATP. They are also involved in other processes like calcium storage, iron compound production, and even influencing cell death and differentiation. Mitochondrial dysfunction can lead to various health issues, including mitochondrial diseases and the development of other conditions. Present in nearly all types of human cell, mitochondria are vital to our survival. [1, 2, 3, 4, 5, 6]

The simple sugar broken down in the mitochondria is glucose. This process, called cellular respiration, occurs in the mitochondria and involves several stages, including the Krebs cycle and the electron transport chain. Glucose is initially broken down in the cytoplasm through glycolysis, producing pyruvate, which then enters the mitochondria for further oxidation, using oxygen to create more energy for proper cell function for life and normal cell death.[14]

Key functions of mitochondria:

- **Energy production:** Mitochondria convert nutrients into ATP, the primary energy currency of the cell. [1, 1, 2, 2, 8]
- **Calcium storage:** They store and regulate calcium ions, important for various cellular processes like muscle contraction and blood clotting. [4, 4, 5, 5]
- **Iron compound production:** Mitochondria produce the iron compound needed for red blood cells to transport oxygen. [4, 4]
- **Cell signaling and differentiation:** They play a role in signaling pathways and cell development. [1, 1, 5, 5]
- **Programmed cell death:** They can trigger cell death pathways when necessary. [5, 5]
- **Regulation of cell cycle and growth:** Mitochondria help control the cell cycle and growth processes. [1, 1]

Mitochondrial structure:

- Mitochondria have an outer and inner membrane, with the inner membrane folded into cristae, which increases surface area for energy production. [1, 6, 9]
- They contain their own DNA, distinct from the cell's nuclear DNA. [1, 1, 10, 10]
- Mitochondria are dynamic and can fuse, divide, and change shape in response to cellular needs. [11, 11, 12, 12]

Mitochondrial diseases:

Mutations in mitochondrial DNA or nuclear genes affecting mitochondrial function can cause mitochondrial diseases.

- These diseases can affect various organs and systems, leading to a range of symptoms like muscle weakness, neurological problems, and heart disease.
- Mitochondrial dysfunction has also been linked to the development of other complex diseases, including cancer and neurodegenerative disorders. [6, 9]

Mitochondrial health:

- Maintaining healthy mitochondria is important for overall health and can be influenced by lifestyle choices like diet and exercise.
- Regular exercise and activity, a diet rich in fruits and vegetables lean proteins. By **avoiding** processed foods, excessive sugar/sugary foods and eliminating environmental toxins, chemicals and of course smoking can help support and improve mitochondrial health. [13]

Cells with higher energy demands, like muscle cells, nerve cells, and liver cells, tend to have more mitochondria than cells with lower energy needs. This is because mitochondria are the primary source of ATP, the cell's energy currency, and cells that perform more work or have higher metabolic rates require more ATP.
Elaboration:

Muscle tissue, especially heart and skeletal muscle, needs a constant supply of energy for contraction and movement. Therefore, these cells have many more mitochondria to produce the ATP needed for these energy-intensive processes, thousands more per cell.

Nerve cells also require a significant amount of energy to transmit signals throughout the body. They have many mitochondria to support the electrochemical processes involved in nerve impulse transmission, hundreds of thousands of mitochondria in nerve bundles and axion

The liver performs numerous metabolic functions, including detoxification and the processing of nutrients. These processes are highly energy-demanding, so liver cells have a large number of mitochondria to generate the necessary ATP.

Sperm cells also have a high number of mitochondria. They require energy to swim to the egg and fertilize it.

] https://en.wikipedia.org/wiki/Mitochondrion
[2] https://my.clevelandclinic.org/health/diseases/15612-mitochondrial-diseases
[3] https://www.khanacademy.org/science/ap-biology/cell-structure-and-function/cell-structures-and-their-functions/v/mitochondria-video
[4] https://www.pfizer.com/news/articles/why_mitochondria_is_the_organelle_of_the_moment
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[12] https://medschool.ucla.edu/research/themed-areas/metabolism-research/mitochondria
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[14] https://courses.lumenlearning.com/suny-ap2/chapter/carbohydrate-metabolism-no-content/

What are mitochondria?

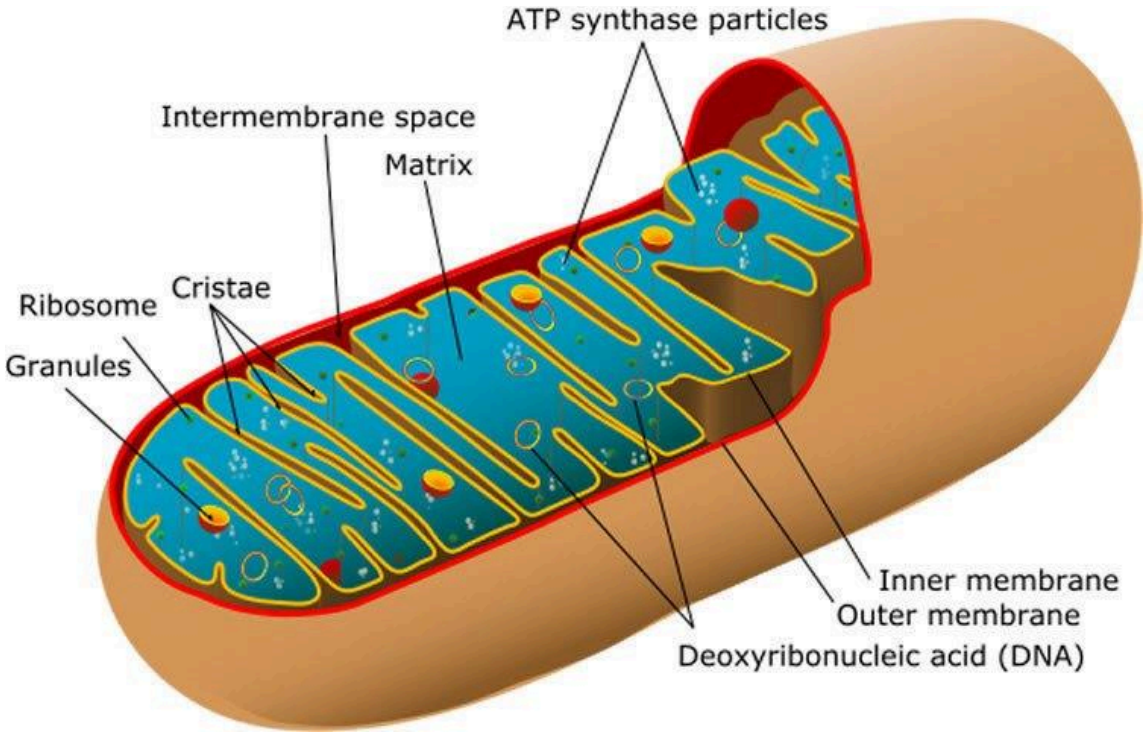
Mitochondria are often referred to as the powerhouses of the cell. Their main function is to generate the energy necessary to power cells. But, there is more to mitochondria than energy production.

Present in nearly all types of human cell, mitochondria are vital to our survival. They generate the majority of our adenosine triphosphate (ATP), the energy currency of the cell.

Mitochondria are also involved in other tasks, such as signaling between cells and cell death, otherwise known as apoptosis.

In this article, we will look at how mitochondria work, what they look like, and explain what happens when they stop doing their job correctly.

The structure of mitochondria



Mitochondria are small, often between 0.75 and 3 micrometers and are not visible under the

microscope unless they are stained.

Unlike other organelles (miniature organs within the cell), they have two membranes, an outer one and an inner one. Each membrane has different functions.

Mitochondria are split into different compartments or regions, each of which carries out distinct roles.

Some of the major regions include the:

Outer membrane: Small molecules can pass freely through the outer membrane. This outer portion includes proteins called porins, which form channels that allow proteins to cross. The outer membrane also hosts a number of enzymes with a wide variety of functions.

Intermembrane space: This is the area between the inner and outer membranes.

Inner membrane: This membrane holds proteins that have several roles. Because there are no porins in the inner membrane, it is impermeable to most molecules. Molecules can only cross the inner membrane in special membrane transporters. The inner membrane is where most ATP is created.

Cristae: These are the folds of the inner membrane. They increase the surface area of the membrane, therefore increasing the space available for chemical reactions.

Matrix: This is the space within the inner membrane. Containing hundreds of enzymes, it is important in the production of ATP. Mitochondrial DNA is housed here (see below).

Different cell types have different numbers of mitochondria. For instance, mature red blood cells have none at all, whereas liver cells can have more than 2,000. Cells with a high demand for energy tend to have greater numbers of mitochondria. Around 40 percent of the cytoplasm in heart muscle cells is taken up by mitochondria.

Although mitochondria are often drawn as oval-shaped organelles, they are constantly dividing (fission) and bonding together (fusion). So, in reality, these organelles are linked together in ever-changing networks.

Also, in sperm cells, the mitochondria are spiraled in the midpiece and provide energy for tail motion.

Mitochondrial DNA

Although most of our DNA is kept in the nucleus of each cell, mitochondria have their own set of DNA. Interestingly, mitochondrial DNA (mtDNA) is more similar to bacterial DNA.

The mtDNA holds the instructions for a number of proteinsTrusted Source and other cellular support equipment across 37 genes.

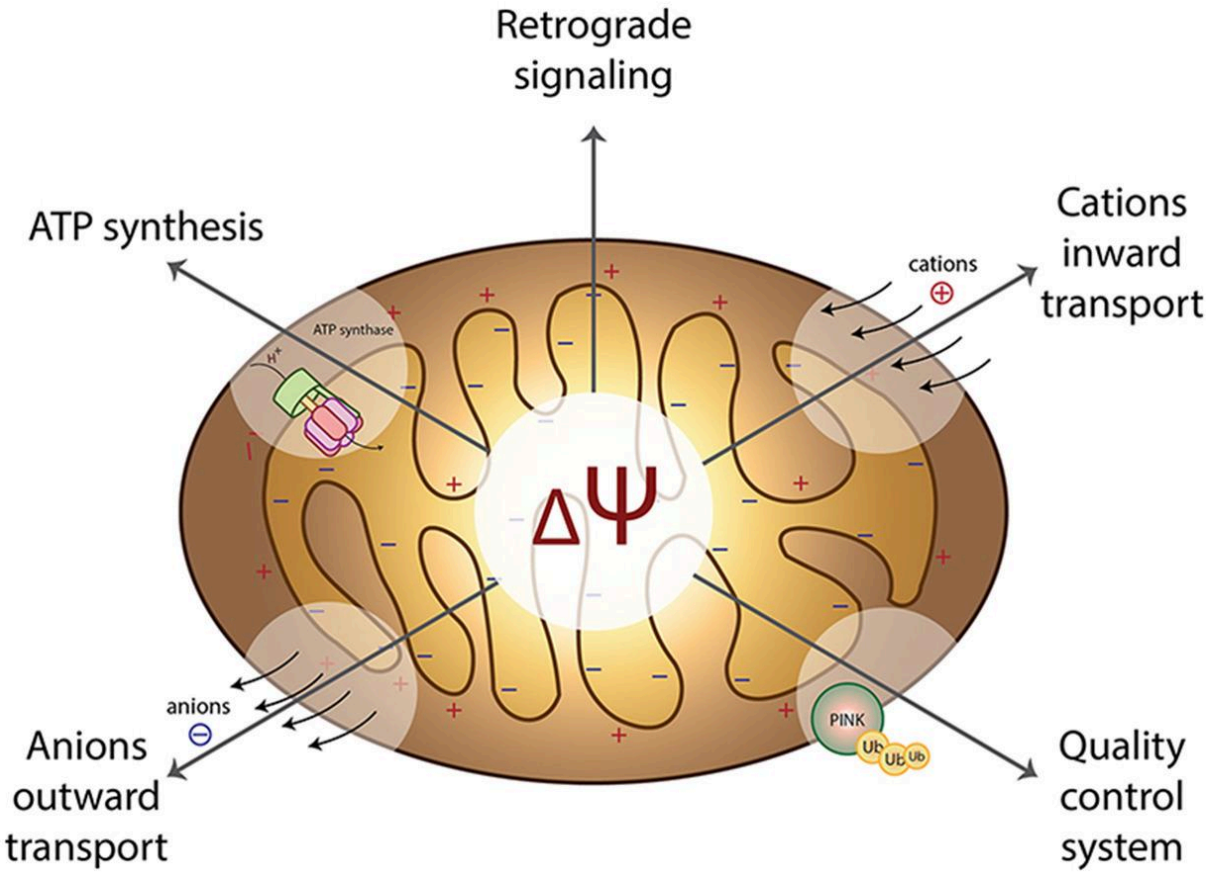
The human genome stored in the nuclei of our cells contains around 3.3 billion base pairs, whereas mtDNA consists of less than 17,000Trusted Source.

During reproduction, half of a child's DNA comes from their father and half from their mother. However,

the child always receives their mtDNA from their mother. Because of this, mtDNA has proven very useful for tracing genetic lines.

For instance, mtDNA analyses have concluded that humans may have originated in Africa relatively recently, around 200,000 years ago, descended from a common ancestor, known as mitochondrial EveTrusted Source.

What do mitochondria do?



Mitochondria are important in a number of processes.

Although the best-known role of mitochondria is energy production, they carry out other important tasks as well.

In fact, only about 3 percent of the genes needed to make a mitochondrion go into its energy production equipment. The vast majority are involved in other jobs that are specific to the cell type where they are found.

Below, we cover a few of the roles of the mitochondria:

Producing energy

ATP, a complex organic chemical found in all forms of life, is often referred to as the molecular unit of currency because it powers metabolic processes. Most ATP is produced in mitochondria through a series of reactions, known as the citric acid cycle or the Krebs cycle.

Energy production mostly takes place on the folds or cristae of the inner membrane.

Mitochondria convert chemical energy from the food we eat into an energy form that the cell can use. This process is called oxidative phosphorylation.

The Krebs cycle produces a chemical called NADH. NADH is used by enzymes embedded in the cristae to produce ATP. In molecules of ATP, energy is stored in the form of chemical bonds. When these chemical bonds are broken, the energy can be used.

Cell death

Cell death, also called apoptosis, is an essential part of life. As cells become old or broken, they are cleared away and destroyed. Mitochondria help decide which cells are destroyed.

Mitochondria release cytochrome C, which activates caspase, one of the chief enzymes involved in destroying cells during apoptosis.

Because certain diseases, such as cancer, involve a breakdown in normal apoptosis, mitochondria are thought to play a role in the disease.

Storing calcium

Calcium is vital for a number of cellular processes. For instance, releasing calcium back into a cell can initiate the release of a neurotransmitter from a nerve cell or hormones from endocrine cells. Calcium is also necessary for muscle function, fertilization, and blood clotting, among other things.

Because calcium is so critical, the cell regulates it tightly. Mitochondria play a part in this by quickly absorbing calcium ions and holding them until they are needed.

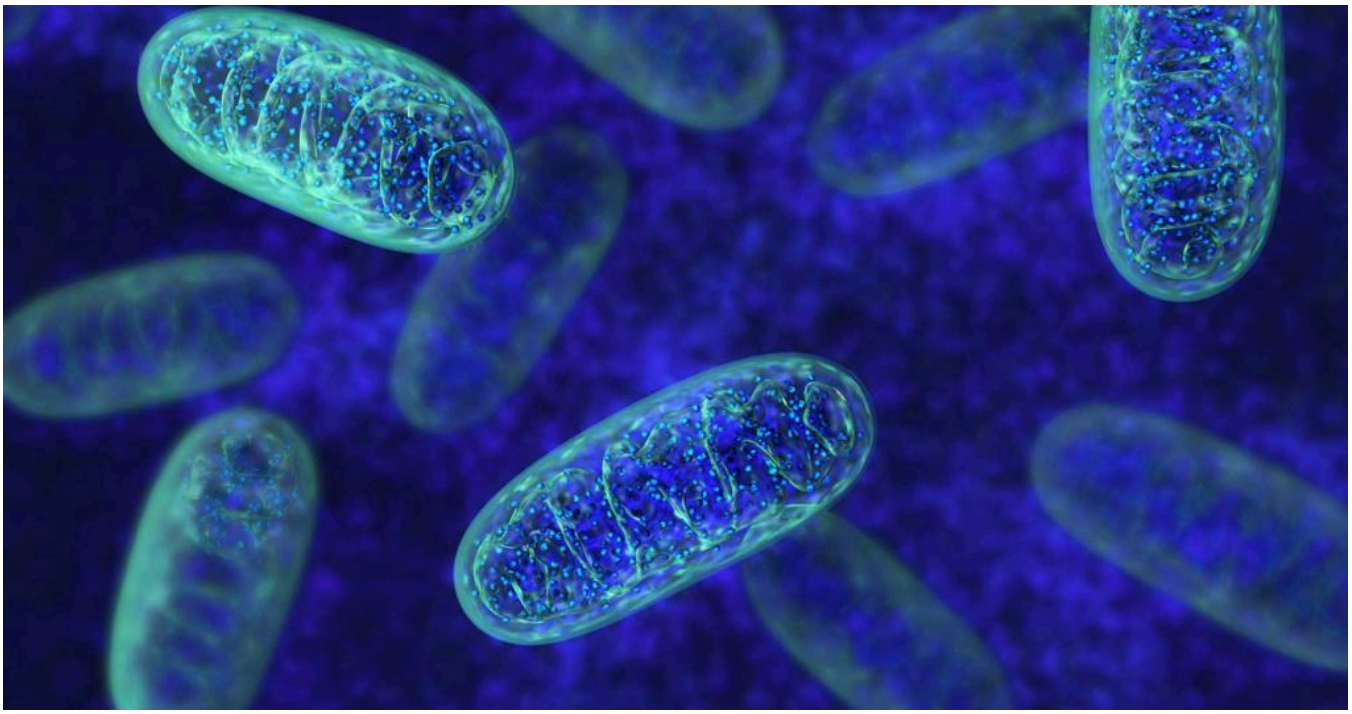
Other roles for calcium in the cell include regulating cellular metabolism, steroid synthesis, and hormone signalingTrusted Source.

Heat production

When we are cold, we shiver to keep warm. But the body can also generate heat in other ways, one of which is by using a tissue called brown fat.

During a process called proton leakTrusted Source, mitochondria can generate heat. This is known as non-shivering thermogenesis. Brown fat is found at its highest levels in babies, when we are more susceptible to cold, and slowly levels reduce as we age.

Mitochondrial disease



If mitochondria do not function correctly, it can cause a range of medical problems.

The DNA within mitochondria is more susceptible to damage than the rest of the genome.

This is because free radicals, which can cause damage to DNA, are produced during ATP synthesis.

Also, mitochondria lack the same protective mechanisms found in the nucleus of the cell.

However, the majority of mitochondrial diseases are due to mutations in nuclear DNA that affect products that end up in the mitochondria. These mutations can either be inherited or spontaneous.

When mitochondria stop functioning, the cell they are in is starved of energy. So, depending on the type of cell, symptoms can vary widely. As a general rule, cells that need the largest amounts of energy, such as heart muscle cells and nerves, are affected the most by faulty mitochondria.

The following passage comes from the United Mitochondrial Disease Foundation:

“Because mitochondria perform so many different functions in different tissues, there are literally hundreds of different mitochondrial diseases. […] Because of the complex interplay between the hundreds of genes and cells that must cooperate to keep our metabolic machinery running smoothly, it is a hallmark of mitochondrial diseases that identical mtDNA mutations may not produce identical diseases.”

Diseases that generate different symptoms but are due to the same mutation are referred to as genocopies.

Conversely, diseases that have the same symptoms but are caused by mutations in different genes are called phenocopies. An example of a phenocopy is Leigh syndrome, which can be caused by several different mutations.

Although symptoms of a mitochondrial disease vary greatly, they might include:

- loss of muscle coordination and weakness
- problems with vision or hearing
- learning disabilities
- heart, liver, or kidney disease
- gastrointestinal problems
- neurological problems, including dementia

Other conditions that are thought to involve some level of mitochondrial dysfunction, include:

- Parkinson's disease
- Alzheimer's disease
- bipolar disorder
- schizophrenia
- chronic fatigue syndrome
- Huntington's disease
- diabetes
- autism

Mitochondria and aging

Over recent years, researchers have investigated a link between mitochondria dysfunction and aging. There are a number of theories surrounding aging, and the mitochondrial free radical theory of aging has become popular over the last decade or so.

The theory is that reactive oxygen species (ROS) are produced in mitochondria, as a byproduct of energy production. These highly charged particles damage DNA, fats, and proteins.

Because of the damage caused by ROS, the functional parts of mitochondria are damaged. When the mitochondria can no longer function so well, more ROS are produced, worsening the damage further.

Although correlations between mitochondrial activity and aging have been found, not all scientists have reached the same conclusions. Their exact role in the aging process is still unknown.

In a nutshell

Mitochondria are, quite possibly, the best-known organelle. And, although they are popularly referred to as the powerhouse of the cell, they carry out a wide range of actions that are much less known about. From calcium storage to heat generation, mitochondria are hugely important to our cells' everyday functions.