Mitochondrial Dna Encyclopedia Article

Mitochondrial Dna

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Mitochondrial Dna

Mitochondria are cellular organelles that generate energy in the form of ATP through oxidative phosphorylation. Each cell contains hundreds of these important organelles. Mitochondria are inherited at conception from the mother through the **cytoplasm** of the egg. The mitochondria, present in all of the cells of the body, are copies of the ones present in at conception in the egg. When cells divide, the mitochondria that are present are randomly distributed to the daughter cells, and the mitochondria themselves then replicate as the cells grow.

Although many of the mitochondrial genes necessary for ATP production and other genes needed by the mitochondria are encoded in the **DNA** of the **chromosomes** in the **nucleus** of the cell, some of the genes expressed in mitochondria are encoded in a small circular chromosome which is contained within the mitochondrion itself. This includes 13 polypeptides, which are components of oxidative phosphorylation **enzymes**, 22 transfer **RNA** (t-RNA) genes, and two genes for ribosomal RNA (r-RNA). Several copies of the mitochondrial chromosome are found in each mitochondrion. These chromosomes are far smaller than the chromosomes found in the nucleus, contain far fewer genes than any of the autosomes, replicate without going through a mitotic cycle, and their morphological structure is more like a bacterial chromosome than it is like the chromosomes found in the nucleus of **eukaryotes**.

Genes which are transmitted through the mitochondrial DNA are inherited exclusively from the mother, since few if any mitochondria are passed along from the sperm. Genetic diseases involving these genes show a distinctive pattern of inheritance in which the trait is passed from an affected female to all of her children. Her daughters will likewise pass the trait on to all of her children, but her sons do not transmit the trait at all.

The types of disorders which are inherited through **mutations** of the mitochondrial DNA tend to involve disorders of nerve function, as neurons require large amounts of energy to function properly. The best known of the mitochondrial disorders is Leber hereditary optic neuropathy (LHON), which involves bilateral central vision loss, which quickly worsens as a result of the death of the optic nerves in early adulthood. Other mitochondrial diseases include Kearns-Sayre syndrome, myoclonus epilepsy with ragged red fibers (MERFF), and mitochondrial encephalomyopathy, lactic acidosis and stroke-like episodes (MELAS).