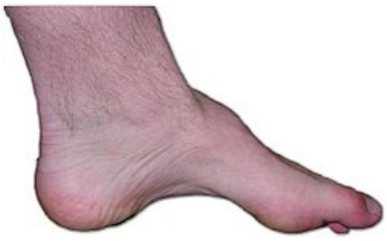
Mitofusin


Manifestation of CMT syndrome

As its name suggests, mitofusin is involved in mitochondrial fusion. It is a membrane protein, that is anchored in the outer membrane of the mitochondria. It has a specific structure divided into several parts. In fact, there are two specific proteins - mitofusin 1 and 2. The gene for **mitofusin 1** (http://omim.org/entry/608506) (MFN1) is located on chromosome 3, the gene for **mitofusin 2** (http://omim.org/entry/608507) (MFN2) on chromosome 1.

In the better studied mitofusin 2, its parts were named **paddle** (tip), **trunk**, **neck** and **terminal GTPase**. According to the proposed model, mitofusins 2 of two mitochondria must fuse, GTP cleavage, conformational change and outer membrane fusion occur. The inner membranes of the mitochondria fuse probably due to the **OPA1** (optic atrophy 1) protein. OPA1 is also a GTPase, named after the syndrome caused by a mutation in its gene.

Thanks to this function, dynamic plasticity of mitochondria can occur inside the cell, as well as remodelling of the mitochondrial network. A mutation in the gene for mitofusin 2 causes a disease called Charcot-Marie-Tooth syndrome (type CMT2A).

Links

Related articles

- Charcot-Marie-Tooth syndrome

External links

- **Database OMIM**
  - http://omim.org/entry/608506 - MFN1
  - http://omim.org/entry/608507 - MFN2
  - http://omim.org/entry/605290 - OPA1
- Wikipedia. *MFN2* [online]. [cit. 2011-04-07]. <https://en.wikipedia.org/wiki/MFN2>.

- https://en.wikipedia.org/wiki/MFN1
- http://www.nature.com/nrn/journal/v9/n7/fig_tab/nrn2417_F2.html
- https://en.wikipedia.org/wiki/OPA1
- https://en.wikipedia.org/wiki/Charcot%E2%80%93Marie%E2%80%93Tooth_disease