

Loss of hepatic LRPPRC alters mitochondrial bioenergetics ...

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May 31, 2017 · Loss of LRPPRC in the liver caused a generalized growth delay, and typical histological features of mitochondrial hepatopathy. At the molecular level, LRPPRC deficiency caused destabilization of polyadenylated mitochondrial mRNAs, altered mitochondrial ultrastructure, and a severe complex IV (CIV) and ATP synthase (CV) assembly defect.

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OXPPOS is carried out in the inner mitochondrial membrane (IMM) by a complex structure denominated respiratory chain (RC). As the term suggests, OXPPOS can be divided into two distinct reactions: an oxidative exergonic pathway called respiration which feeds a second pathway consisting of phosphorylation of ADP into ATP.



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(PDF) Liver Disease in Mitochondrial Disordershttps://www.researchgate.net/publication/6156517_Liver_Disease_in_Mitochondrial_Disorders

A) Photomicrograph of liver biopsy from a 3-month-old child with POLG mutations and mitochondrial DNA depletion syndrome, showing microvesicular steatosis, cholestasis with bile pigment in ...

Gastrointestinal manifestations of mitochondrial disorders: a systematic review - Josef Finsterer, Marlies Frank, 2017<https://journals.sagepub.com/doi/full/10.1177/1756283X16666806>

Mitochondrial disorders (MIDs) due to respiratory-chain defects or nonrespiratory chain defects are usually multisystem conditions [mitochondrial multiorgan disorder syndrome (MIMODS)] affecting the central ne...

Name of Journal: *World Journal of Hepatology*

Manuscript NO: 68483

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Mitochondrial hepatopathy: Respiratory chain disorders- 'breathing in and out of the liver'

Gopan A *et al.* Mitochondrial Hepatopathy: Respiratory chain disorders

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Jul 18, 2005 · **Mitochondrial respiratory chain** is the result of the interplay of two physically and functionally separated genomes, the nuclear DNA and the mtDNA. Human mtDNA is a 16.6 kb circular double-stranded DNA containing only 37 genes.

Cited by: 145

Author: Antonella Spinazzola, Massimo Zeviani

Publish Year: 2005

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