Loss of hepatic LRPPRC alters mitochondrial bioenergetics...
https://academic.oup.com/hmg/article/26/16/3186/3859089
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 (CIV) assembly defect.
Cited by: 20  Author: Alexanne Cuillerier, Shamisa Honarmand, v...
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europepmc.org/abstract/MED/28575497
Aug 01, 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 (CIV) assembly defect.
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Disorders from perturbations of nuclear-mitochondrial...
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OXPHOS is carried out in the inner mitochondrial membrane (IMM) by a complex structure denominated respiratory chain (RC). As the term suggests, OXPHOS 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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Amit Mitochondrial respiratory chain disorder (MRCDD) can cause acute liver failure (ALF), which may necessitate liver transplantation (LT). However, MRCDD is often difficult to diagnose before LT ...

Metabolic disorders of fetal life: Glycogenoses and mitochondrial defects of the mitochondrial respiratory chain ...

First, in fetal tissues anaerobic glycolysis is the major source of cellular ATP; 52 thus possibly explaining any fetal wastage due to respiratory chain disorders is not noted as a major feature in the three retrospe.

(PDF) Liver Disease in Mitochondrial Disorders
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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
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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...
None of Journals: World Journal of Hepatology
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Manuscript Type: MINIREVIEW

Mitochondrial myopathy: Respiratory chain disorders—"breathing in and out of the loop"

Gopas A et al. Mitochondrial Myopathy: Respiratory chain disorders
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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 (CV) and ATP synthase (CV) assembly defect.
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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.
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Liver Hemosiderosis - an overview | ScienceDirect Topics
https://www.sciencedirect.com/topics/medicine-and-dentistry/liver-hemosiderosis
The usual clinical features are either severe neonatal hepatitis syndrome or else neonatal liver failure. 1021–1028 Children with mitochondrial respiratory chain defects may be at increased risk of developing hepatocellular carcinoma. 1029 A cytochrome c oxidase deficiency found in Quebec kindreds may be associated with fatty liver. 1030...