

CPT1A,B transfers PALM to CAR

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Introduction

Reactome is open-source, open access, manually curated and peer-reviewed pathway database. Pathway annotations are authored by expert biologists, in collaboration with Reactome editorial staff and cross-referenced to many bioinformatics databases. A system of evidence tracking ensures that all assertions are backed up by the primary literature. Reactome is used by clinicians, geneticists, genomics researchers, and molecular biologists to interpret the results of high-throughput experimental studies, by bioinformaticians seeking to develop novel algorithms for mining knowledge from genomic studies, and by systems biologists building predictive models of normal and disease variant pathways.

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Literature references

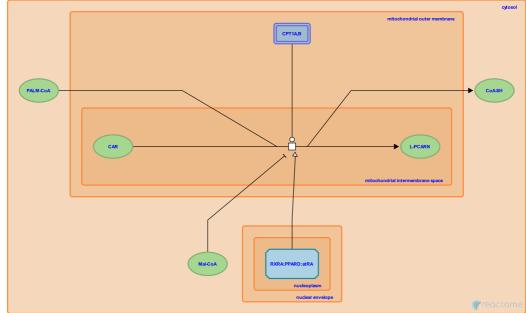
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This document contains 1 reaction (see Table of Contents)

CPT1A,B transfers PALM to CAR ↗

Stable identifier: R-HSA-200406

Type: transition



Compartments: cytosol, mitochondrial outer membrane, mitochondrial intermembrane space

Carnitine palmitoyl transferase 1 (CPT1) associated with the inner mitochondrial membrane, catalyzes the reaction of palmitoyl-CoA (PALM-CoA) from the cytosol with carnitine (CAR) in the mitochondrial intermembrane space to form palmitoylcarnitine (L-PCARN) and CoA-SH. Three CPT1 isoforms exist; CPT1A, B and C. In the body, CPT1A is most abundant in liver while CPT1B is abundant in muscle. CPT1C is mainly expressed in neurons and localises to the ER and not to the mitochondria. It has little or no enzymatic activity in fatty acid oxidation. Both CPT1A and CPT1B are inhibited by malonyl-CoA (Morillas et al. 2002, 2004; Zammit et al. 2001; Zhu et al. 1997). Mutations in CPT1A are associated with defects in fatty acid metabolism and fasting intolerance, consistent with the role assigned to CPT1 from studies in vitro and in animal models (IJIst et al. 1998; Gobin et al. 2003). In the nucleus, cellular retinoic acid-binding protein 1 or 2 (CRABP1 or 2), bound to all-trans-retinoic acid (atRA), directly binds to the heterodimeric complex of retinoic acid receptor alpha RXRA) and peroxisome proliferator-activated receptor delta (PPARD). When bound to PPARD, atRA can significantly increase the expression of proteins involved in fatty acid oxidation such as CPT1A via its induction of PPARD (Amengual et al. 2012).

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Editions

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