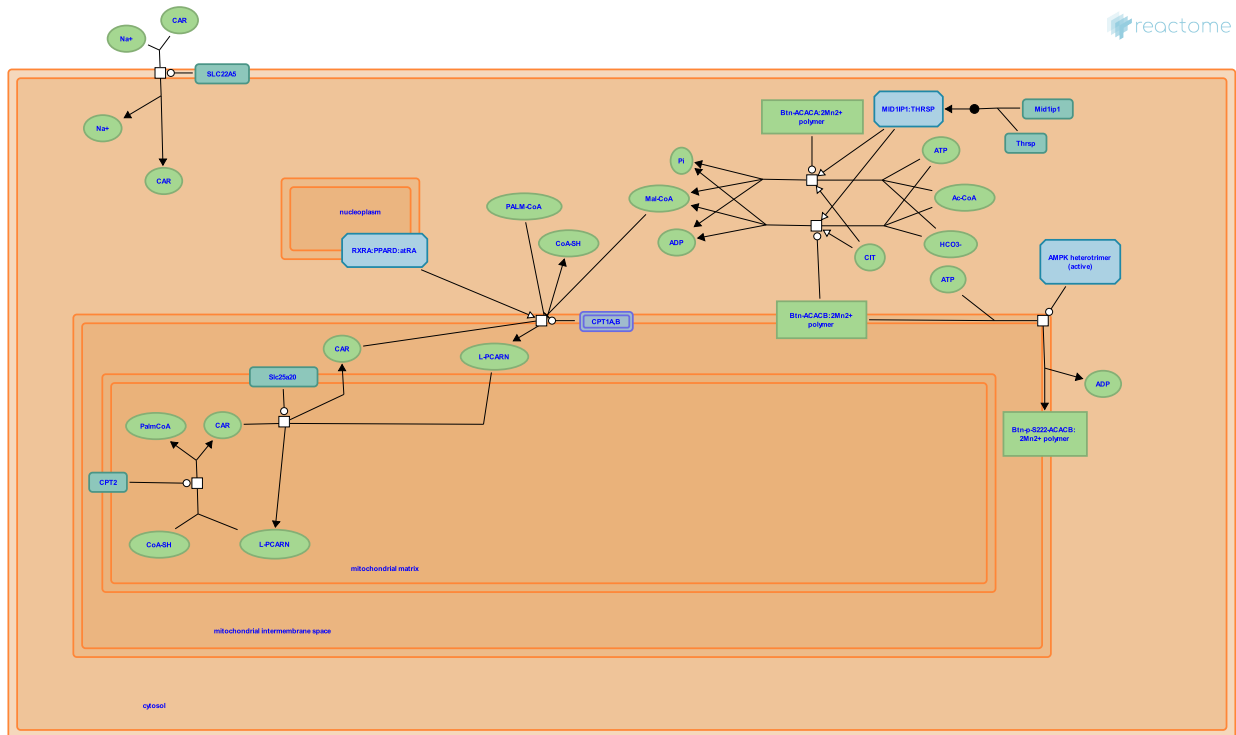


# Carnitine metabolism



European Bioinformatics Institute, New York University Langone Medical Center, Ontario Institute for Cancer Research, Oregon Health and Science University.

The contents of this document may be freely copied and distributed in any media, provided the authors, plus the institutions, are credited, as stated under the terms of [Creative Commons Attribution 4.0 International \(CC BY 4.0\) License](https://creativecommons.org/licenses/by/4.0/). For more information see our [license](#).

This is just an excerpt of a full-length report for this pathway. To access the complete report, please download it at the [Reactome Textbook](#).

03/05/2024

## 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.

The development of Reactome is supported by grants from the US National Institutes of Health (P41 HG003751), University of Toronto (CFREF Medicine by Design), European Union (EU STRP, EMI-CD), and the European Molecular Biology Laboratory (EBI Industry program).

## Literature references

- Fabregat, A., Sidiropoulos, K., Viteri, G., Forner, O., Marin-Garcia, P., Arnau, V. et al. (2017). Reactome pathway analysis: a high-performance in-memory approach. *BMC bioinformatics*, 18, 142. [↗](#)
- Sidiropoulos, K., Viteri, G., Sevilla, C., Jupe, S., Webber, M., Orlic-Milacic, M. et al. (2017). Reactome enhanced pathway visualization. *Bioinformatics*, 33, 3461-3467. [↗](#)
- Fabregat, A., Jupe, S., Matthews, L., Sidiropoulos, K., Gillespie, M., Garapati, P. et al. (2018). The Reactome Pathway Knowledgebase. *Nucleic Acids Res*, 46, D649-D655. [↗](#)
- Fabregat, A., Korninger, F., Viteri, G., Sidiropoulos, K., Marin-Garcia, P., Ping, P. et al. (2018). Reactome graph database: Efficient access to complex pathway data. *PLoS computational biology*, 14, e1005968. [↗](#)

Reactome database release: 88

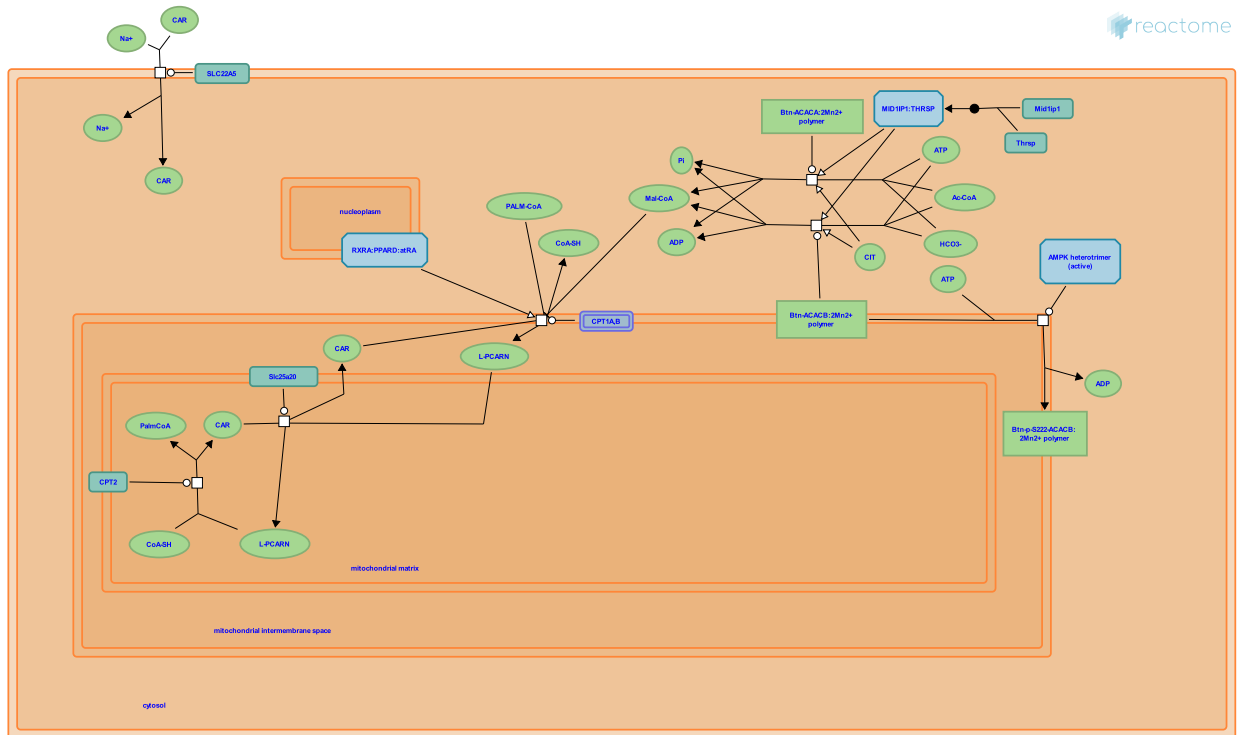
This document contains 1 pathway and 8 reactions ([see Table of Contents](#))

## Carnitine metabolism ↗

**Stable identifier:** R-MMU-200425

**Compartments:** mitochondrion, cytosol

**Inferred from:** Carnitine metabolism (Homo sapiens)



This event has been computationally inferred from an event that has been demonstrated in another species.

The inference is based on the homology mapping from PANTHER. Briefly, reactions for which all involved PhysicalEntities (in input, output and catalyst) have a mapped orthologue/paralogue (for complexes at least 75% of components must have a mapping) are inferred to the other species. High level events are also inferred for these events to allow for easier navigation.

[More details and caveats of the event inference in Reactome.](http://www.pantherdb.org/about.jsp) For details on PANTHER see also: <http://www.pantherdb.org/about.jsp>

## OCTN2 / SLC22A5 transports CAR from extracellular space to cytosol ↗

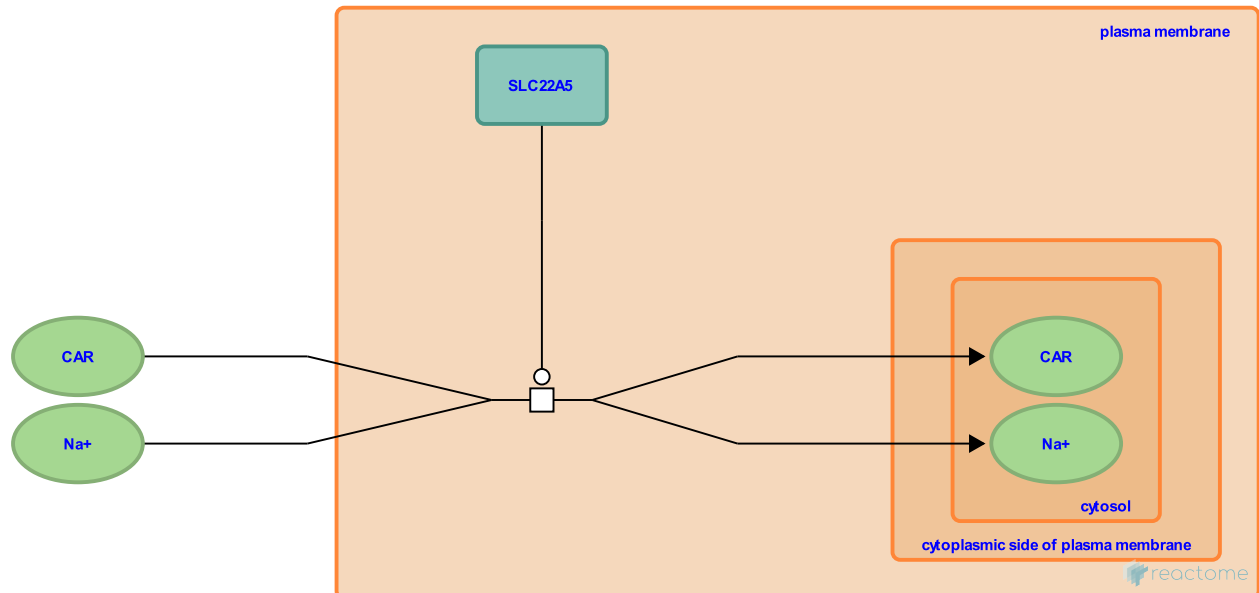
**Location:** [Carnitine metabolism](#)

**Stable identifier:** R-MMU-165026

**Type:** transition

**Compartments:** plasma membrane

**Inferred from:** [OCTN2 / SLC22A5 transports CAR from extracellular space to cytosol \(Homo sapiens\)](#)



This event has been computationally inferred from an event that has been demonstrated in another species.

The inference is based on the homology mapping from PANTHER. Briefly, reactions for which all involved PhysicalEntities (in input, output and catalyst) have a mapped orthologue/paralogue (for complexes at least 75% of components must have a mapping) are inferred to the other species. High level events are also inferred for these events to allow for easier navigation.

[More details and caveats of the event inference in Reactome.](#) For details on PANTHER see also: <http://www.pantherdb.org/about.jsp>

## CPT1A,B transfers PALM to CAR ↗

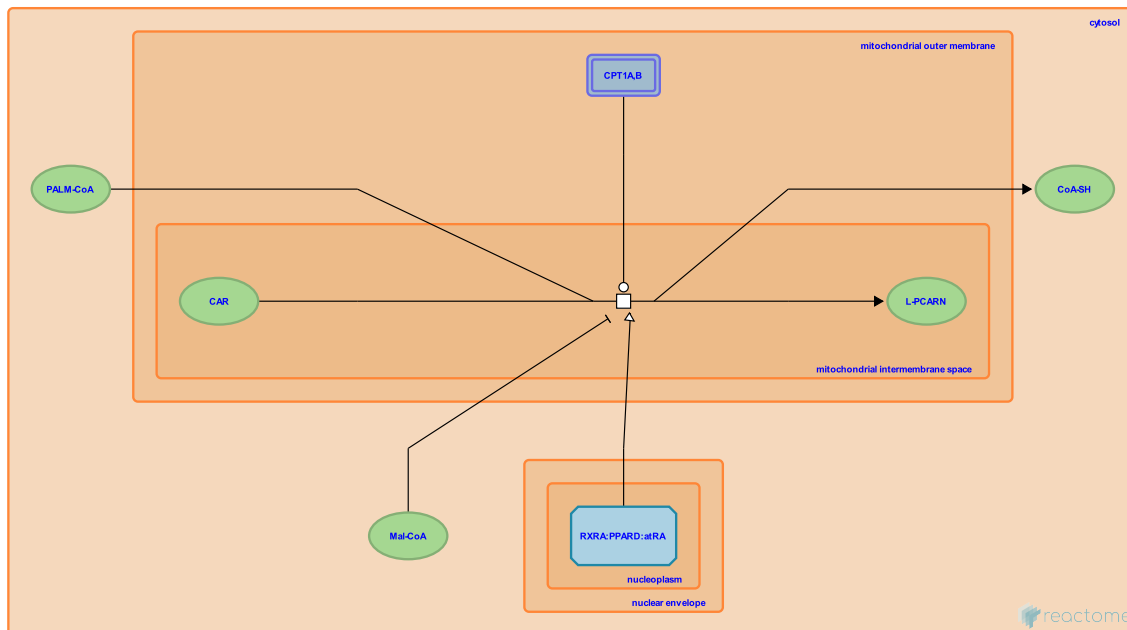
**Location:** Carnitine metabolism

**Stable identifier:** R-MMU-200406

**Type:** transition

**Compartments:** mitochondrial intermembrane space, cytosol, mitochondrial outer membrane

**Inferred from:** CPT1A,B transfers PALM to CAR (Homo sapiens)



This event has been computationally inferred from an event that has been demonstrated in another species.

The inference is based on the homology mapping from PANTHER. Briefly, reactions for which all involved PhysicalEntities (in input, output and catalyst) have a mapped orthologue/paralogue (for complexes at least 75% of components must have a mapping) are inferred to the other species. High level events are also inferred for these events to allow for easier navigation.

[More details and caveats of the event inference in Reactome.](http://www.pantherdb.org/about.jsp) For details on PANTHER see also: <http://www.pantherdb.org/about.jsp>

**Followed by:** Exchange of palmitoylcarnitine and carnitine across the inner mitochondrial membrane

## Exchange of palmitoylcarnitine and carnitine across the inner mitochondrial membrane ↗

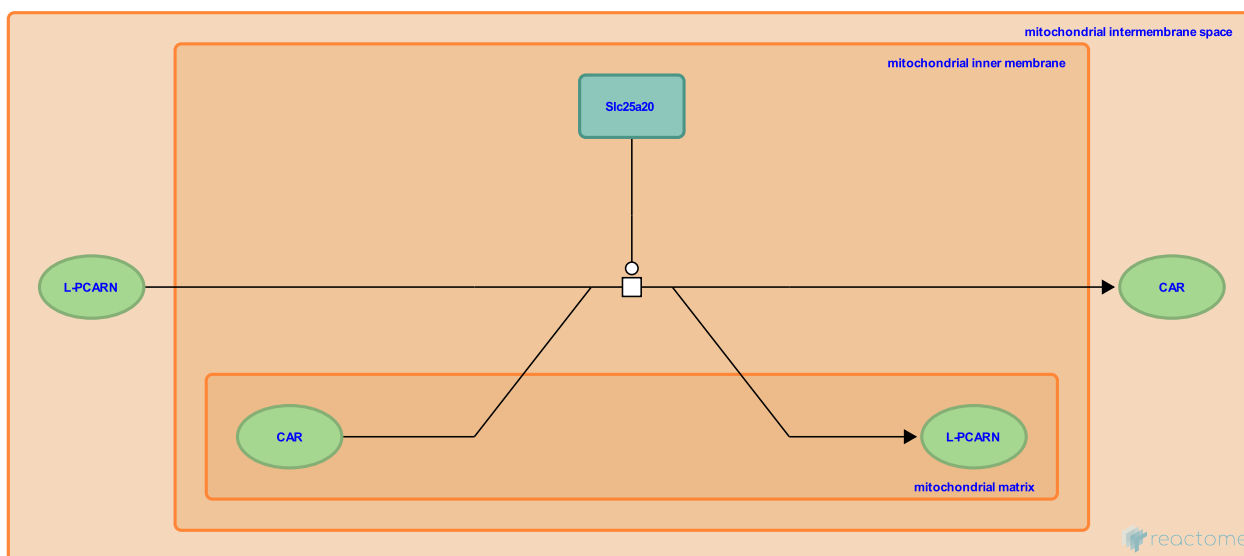
**Location:** [Carnitine metabolism](#)

**Stable identifier:** R-MMU-200424

**Type:** transition

**Compartments:** mitochondrial inner membrane, mitochondrial intermembrane space, mitochondrial matrix

**Inferred from:** [Exchange of palmitoylcarnitine and carnitine across the inner mitochondrial membrane \(Homo sapiens\)](#)



This event has been computationally inferred from an event that has been demonstrated in another species.

The inference is based on the homology mapping from PANTHER. Briefly, reactions for which all involved PhysicalEntities (in input, output and catalyst) have a mapped orthologue/paralogue (for complexes at least 75% of components must have a mapping) are inferred to the other species. High level events are also inferred for these events to allow for easier navigation.

[More details and caveats of the event inference in Reactome.](#) For details on PANTHER see also: <http://www.pantherdb.org/about.jsp>

**Preceded by:** [CPT1A,B transfers PALM to CAR](#)

**Followed by:** [palmitoylcarnitine + CoASH => palmitoyl-CoA + carnitine](#)

**palmitoylcarnitine + CoASH => palmitoyl-CoA + carnitine** ↗

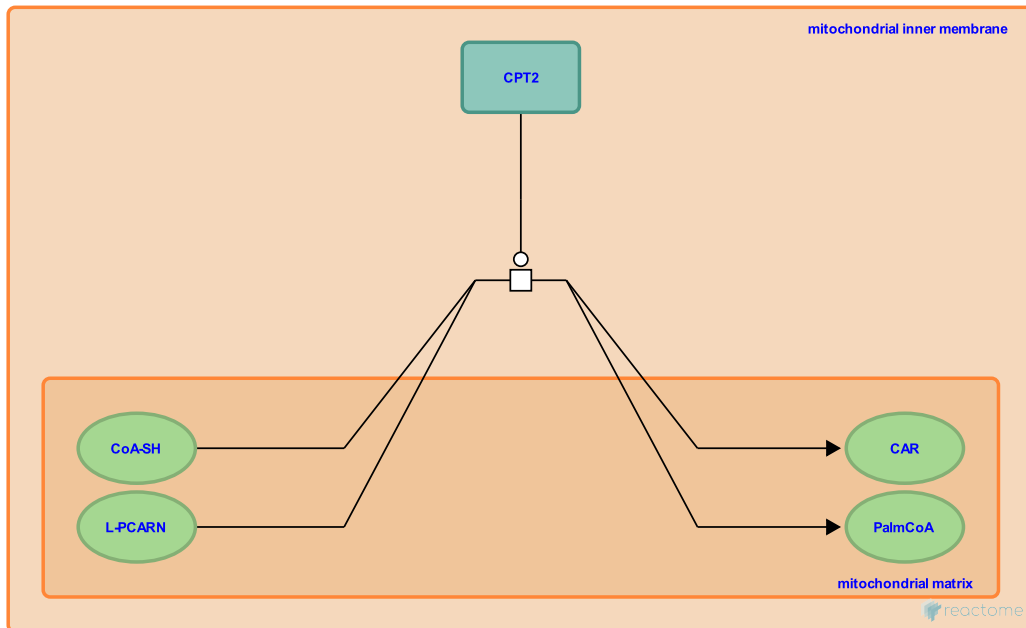
**Location:** Carnitine metabolism

**Stable identifier:** R-MMU-200410

**Type:** transition

**Compartments:** mitochondrial inner membrane, mitochondrial matrix

**Inferred from:** palmitoylcarnitine + CoASH => palmitoyl-CoA + carnitine (Homo sapiens)



This event has been computationally inferred from an event that has been demonstrated in another species.

The inference is based on the homology mapping from PANTHER. Briefly, reactions for which all involved PhysicalEntities (in input, output and catalyst) have a mapped orthologue/paralogue (for complexes at least 75% of components must have a mapping) are inferred to the other species. High level events are also inferred for these events to allow for easier navigation.

[More details and caveats of the event inference in Reactome.](http://www.pantherdb.org/about.jsp) For details on PANTHER see also: <http://www.pantherdb.org/about.jsp>

**Preceded by:** Exchange of palmitoylcarnitine and carnitine across the inner mitochondrial membrane

## MID1IP1 binds THRSP ↗

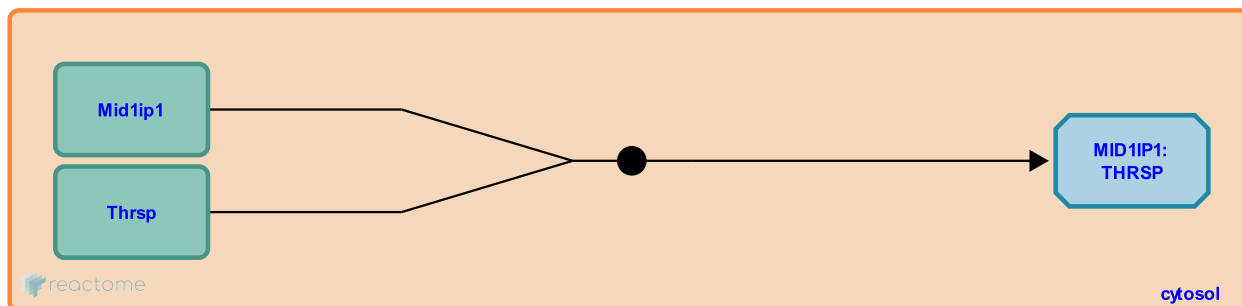
**Location:** [Carnitine metabolism](#)

**Stable identifier:** R-MMU-8866941

**Type:** binding

**Compartments:** cytosol

**Inferred from:** [MID1IP1 binds THRSP \(Homo sapiens\)](#)



This event has been computationally inferred from an event that has been demonstrated in another species.

The inference is based on the homology mapping from PANTHER. Briefly, reactions for which all involved PhysicalEntities (in input, output and catalyst) have a mapped orthologue/paralogue (for complexes at least 75% of components must have a mapping) are inferred to the other species. High level events are also inferred for these events to allow for easier navigation.

[More details and caveats of the event inference in Reactome.](#) For details on PANTHER see also: <http://www.pantherdb.org/about.jsp>



## Btn-ACACA:2Mn2+ polymer carboxylates Ac-CoA to form Mal-CoA ↗

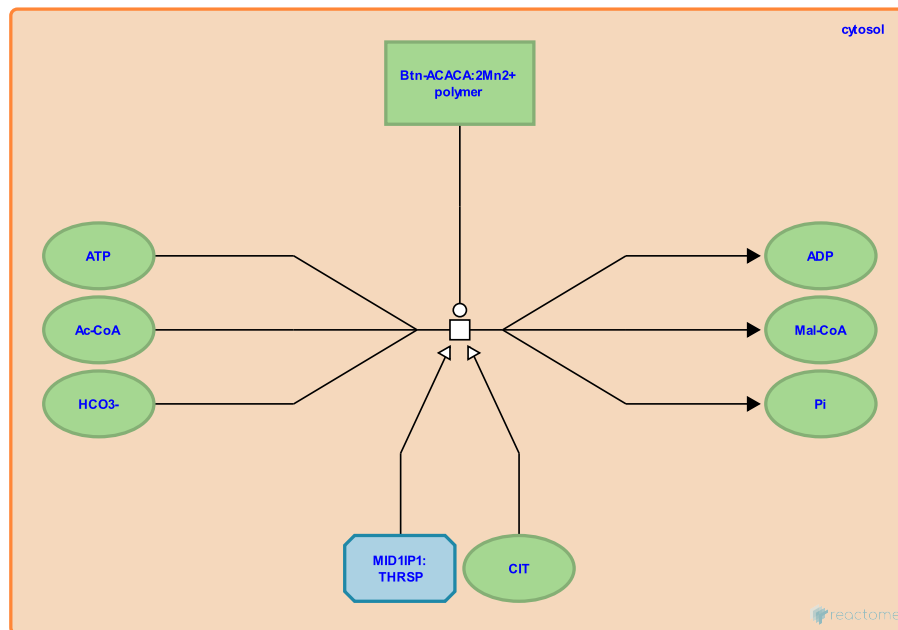
**Location:** Carnitine metabolism

**Stable identifier:** R-MMU-75851

**Type:** transition

**Compartments:** cytosol

**Inferred from:** Btn-ACACA:2Mn2+ polymer carboxylates Ac-CoA to form Mal-CoA (Homo sapiens)



This event has been computationally inferred from an event that has been demonstrated in another species.

The inference is based on the homology mapping from PANTHER. Briefly, reactions for which all involved PhysicalEntities (in input, output and catalyst) have a mapped orthologue/paralogue (for complexes at least 75% of components must have a mapping) are inferred to the other species. High level events are also inferred for these events to allow for easier navigation.

[More details and caveats of the event inference in Reactome.](http://www.pantherdb.org/about.jsp) For details on PANTHER see also: <http://www.pantherdb.org/about.jsp>

## Btn-ACACB:2Mn2+ polymer carboxylates Ac-CoA to form Mal-CoA ↗

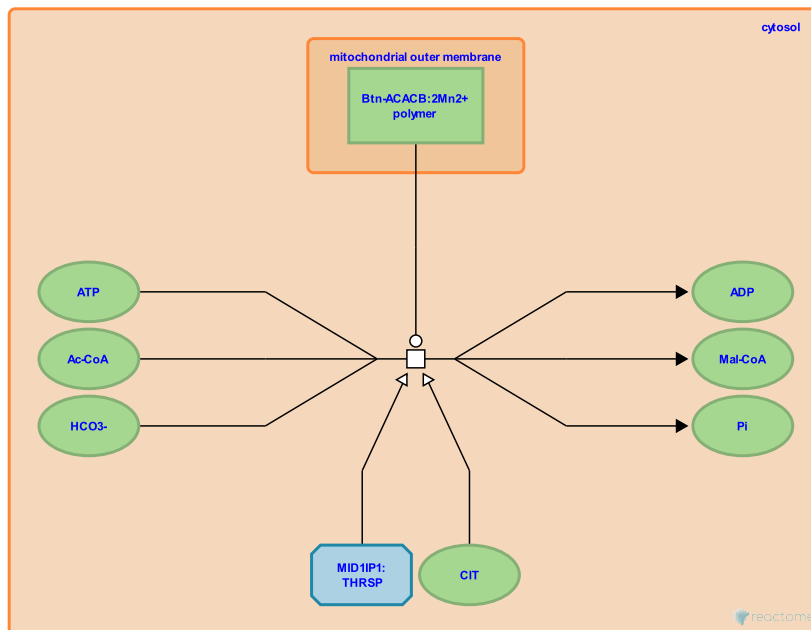
**Location:** Carnitine metabolism

**Stable identifier:** R-MMU-8876889

**Type:** transition

**Compartments:** cytosol, mitochondrial outer membrane

**Inferred from:** Btn-ACACB:2Mn2+ polymer carboxylates Ac-CoA to form Mal-CoA (Homo sapiens)



This event has been computationally inferred from an event that has been demonstrated in another species.

The inference is based on the homology mapping from PANTHER. Briefly, reactions for which all involved PhysicalEntities (in input, output and catalyst) have a mapped orthologue/paralogue (for complexes at least 75% of components must have a mapping) are inferred to the other species. High level events are also inferred for these events to allow for easier navigation.

[More details and caveats of the event inference in Reactome.](http://www.pantherdb.org/about.jsp) For details on PANTHER see also: <http://www.pantherdb.org/about.jsp>

## pAMPK inactivates ACACB, inhibiting malonyl-CoA synthesis ↗

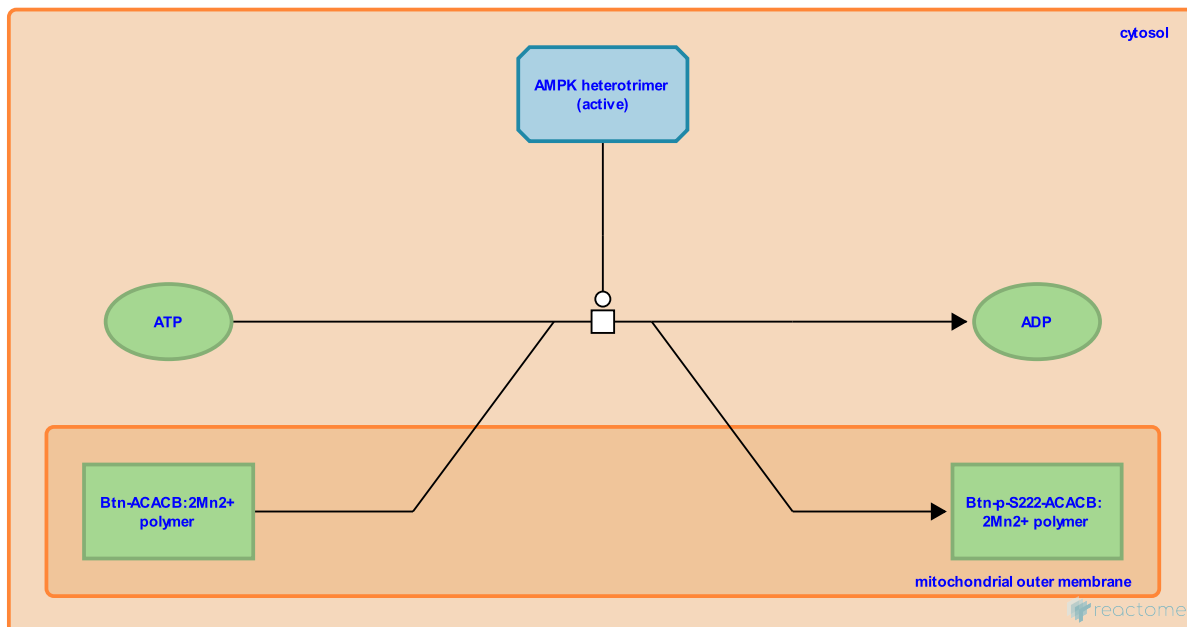
**Location:** [Carnitine metabolism](#)

**Stable identifier:** R-MMU-200423

**Type:** transition

**Compartments:** cytosol, mitochondrial outer membrane

**Inferred from:** [pAMPK inactivates ACACB, inhibiting malonyl-CoA synthesis \(Homo sapiens\)](#)



This event has been computationally inferred from an event that has been demonstrated in another species.

The inference is based on the homology mapping from PANTHER. Briefly, reactions for which all involved PhysicalEntities (in input, output and catalyst) have a mapped orthologue/paralogue (for complexes at least 75% of components must have a mapping) are inferred to the other species. High level events are also inferred for these events to allow for easier navigation.

[More details and caveats of the event inference in Reactome.](#) For details on PANTHER see also: <http://www.pantherdb.org/about.jsp>

# Table of Contents

Introduction	1
☒ Carnitine metabolism	2
↳ OCTN2 / SLC22A5 transports CAR from extracellular space to cytosol	3
↳ CPT1A,B transfers PALM to CAR	4
↳ Exchange of palmitoylcarnitine and carnitine across the inner mitochondrial membrane	5
↳ palmitoylcarnitine + CoASH => palmitoyl-CoA + carnitine	6
↳ MID1IP1 binds THRSP	7
↳ Btn-ACACA:2Mn <sup>2+</sup> polymer carboxylates Ac-CoA to form Mal-CoA	8
↳ Btn-ACACB:2Mn <sup>2+</sup> polymer carboxylates Ac-CoA to form Mal-CoA	9
↳ pAMPK inactivates ACACB, inhibiting malonyl-CoA synthesis	10
Table of Contents	11