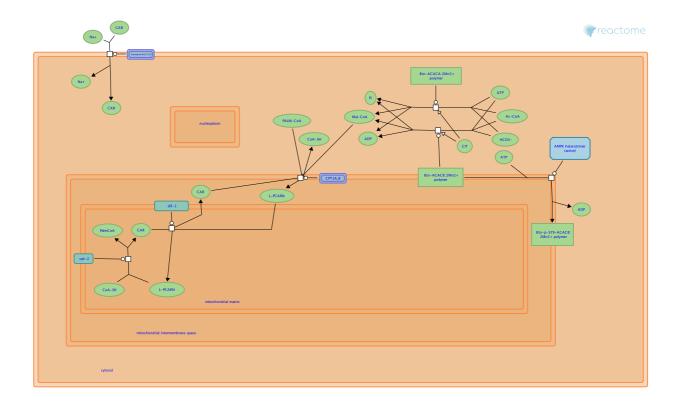


Carnitine metabolism



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

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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

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 data-base: Efficient access to complex pathway data. *PLoS computational biology, 14*, e1005968.

Reactome database release: 77

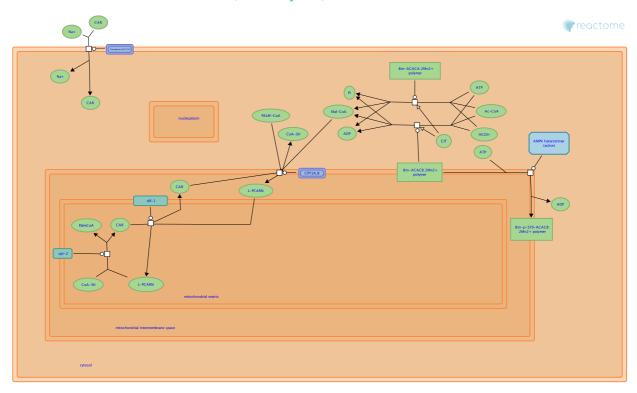
This document contains 1 pathway and 7 reactions (see Table of Contents)

Carnitine metabolism

Stable identifier: R-CEL-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. For details on PANTHER see also: http://www.pantherdb.org/about.jsp

OCTN2 / SLC22A5 transports CAR from extracellular space to cytosol 7

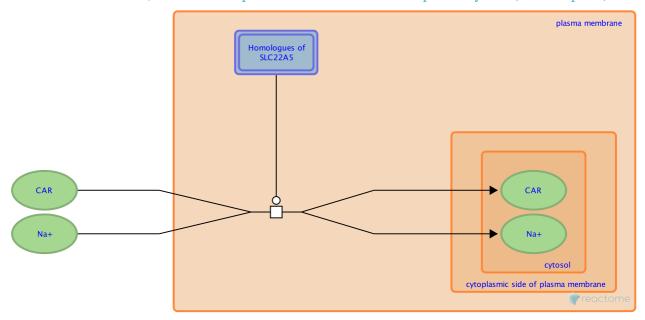
Location: Carnitine metabolism

Stable identifier: R-CEL-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.

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CPT1A,B transfers PALM to CAR 对

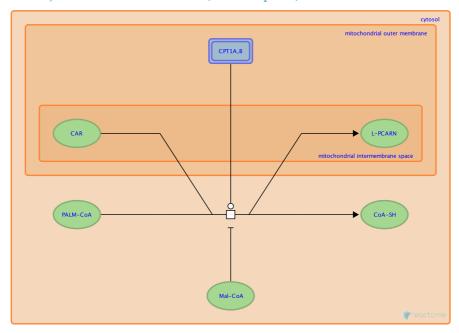
Location: Carnitine metabolism

Stable identifier: R-CEL-200406

Type: transition

Compartments: cytosol, mitochondrial outer membrane, mitochondrial intermembrane space

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.

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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

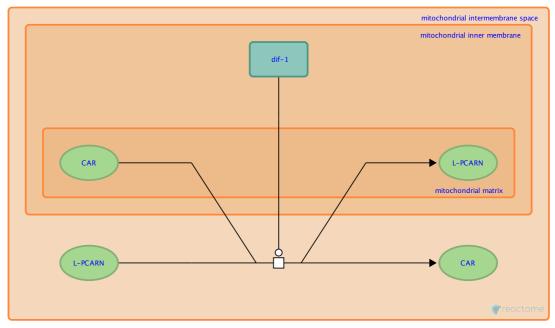
Stable identifier: R-CEL-200424

Type: transition

Compartments: mitochondrial intermembrane space, mitochondrial inner membrane, 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.

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Preceded by: CPT1A,B transfers PALM to CAR

Followed by: palmitoylcarnitine + CoASH => palmitoyl-CoA + carnitine

palmitoylcarnitine + CoASH => palmitoyl-CoA + carnitine 7

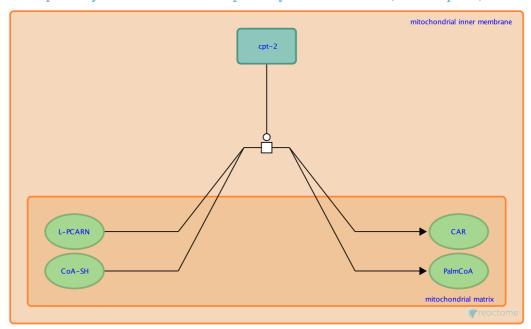
Location: Carnitine metabolism

Stable identifier: R-CEL-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.

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

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

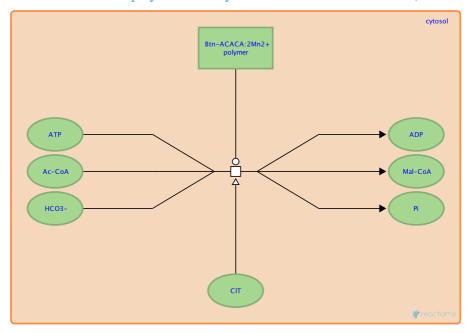
Location: Carnitine metabolism

Stable identifier: R-CEL-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.

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

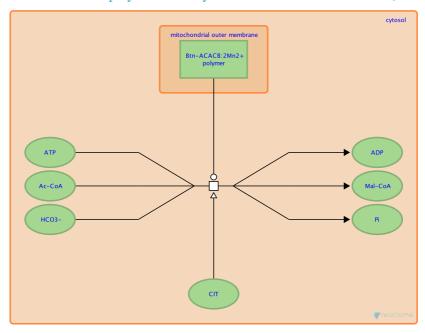
Location: Carnitine metabolism

Stable identifier: R-CEL-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.

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pAMPK inactivates ACACB, inhibiting malonyl-CoA synthesis

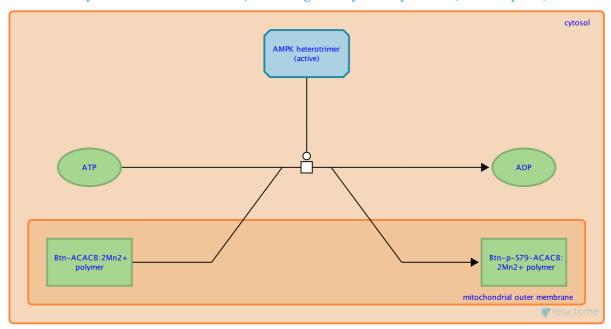
Location: Carnitine metabolism

Stable identifier: R-CEL-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

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