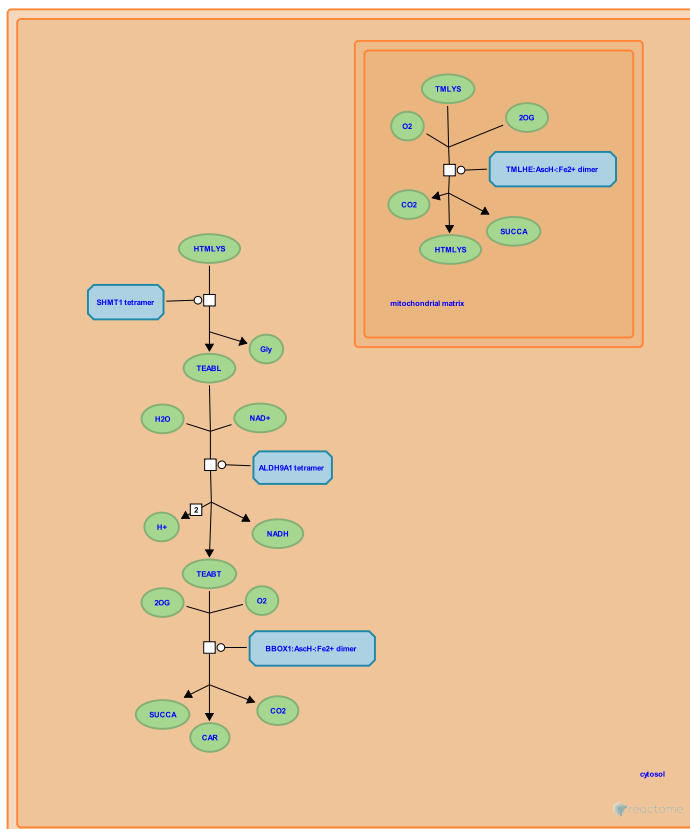


Carnitine synthesis



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](https://reactome.org/About-Reactome/Reactome-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](https://reactome.org/About-Reactome/Reactome-Textbook).

04/05/2024

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

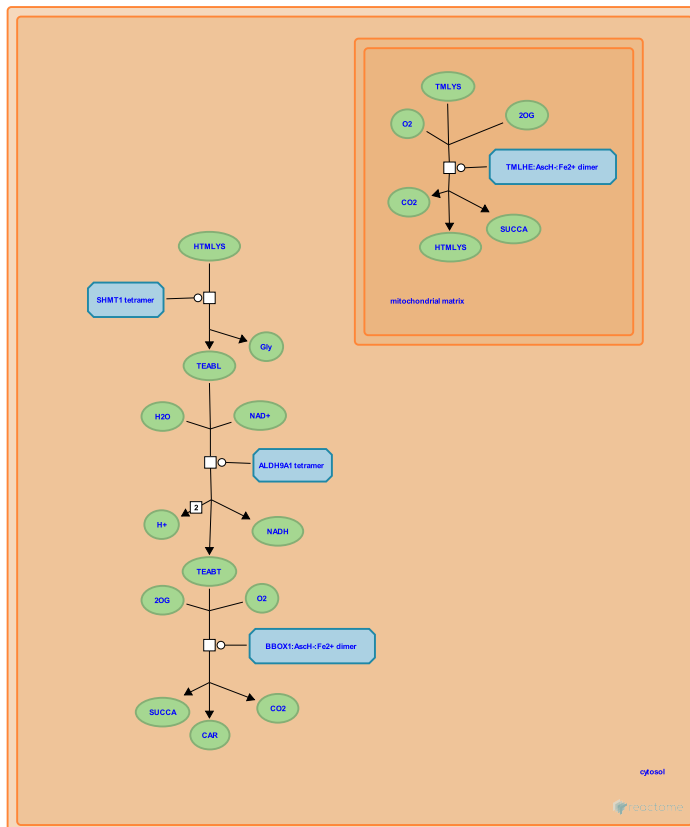
This document contains 1 pathway and 4 reactions ([see Table of Contents](#))

Carnitine synthesis ↗

Stable identifier: R-CFA-71262

Compartments: cytosol, mitochondrial matrix

Inferred from: Carnitine 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.](http://www.pantherdb.org/about.jsp) For details on PANTHER see also: <http://www.pantherdb.org/about.jsp>

TMLHE dimer dioxygenates TMLYS and 2OG to form HTMLYS and SUCCA ↗

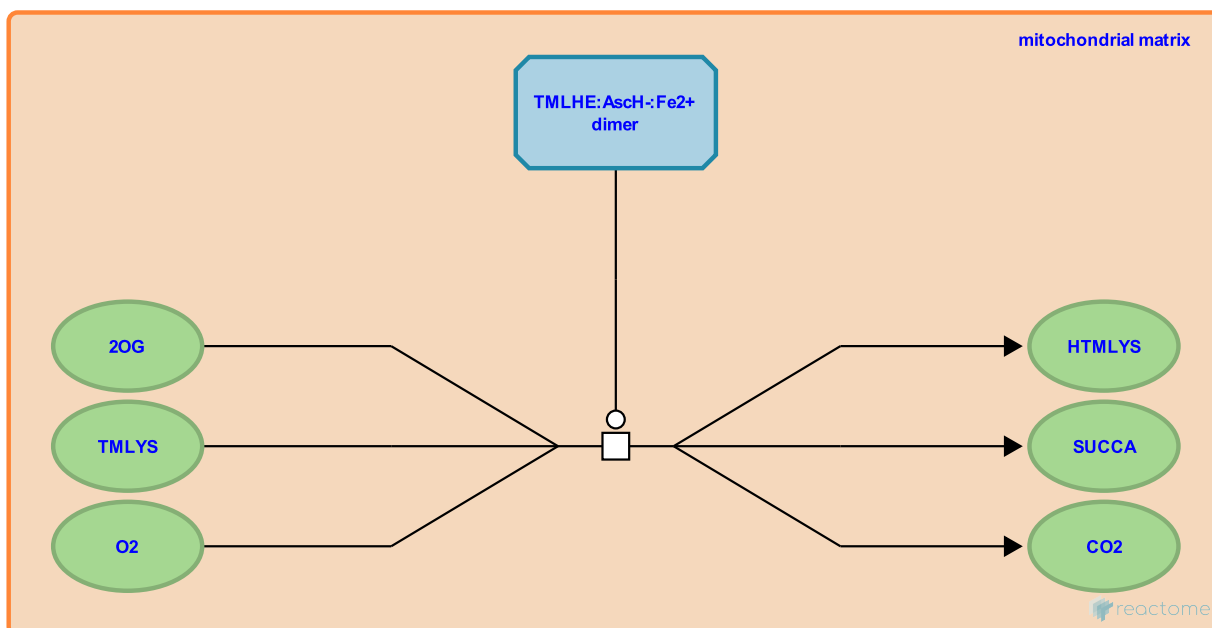
Location: [Carnitine synthesis](#)

Stable identifier: R-CFA-71241

Type: transition

Compartments: mitochondrial matrix

Inferred from: [TMLHE dimer dioxygenates TMLYS and 2OG to form HTMLYS and SUCCA \(Homo sapiens\)](#)



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>

SHMT1 tetramer cleaves HTMLYS to yield TEABL and Gly ↗

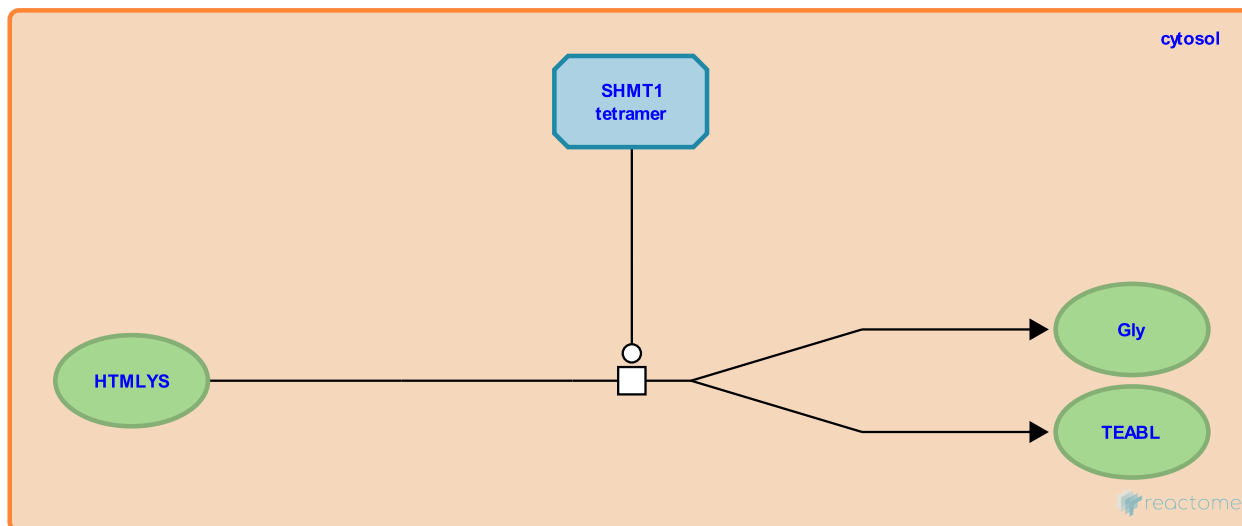
Location: [Carnitine synthesis](#)

Stable identifier: R-CFA-71249

Type: transition

Compartments: cytosol

Inferred from: [SHMT1 tetramer cleaves HTMLYS to yield TEABL and Gly \(Homo sapiens\)](#)



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>

Followed by: [ALDH9A1 tetramer dehydrogenates TEABL to form TEABT](#)

ALDH9A1 tetramer dehydrogenates TEABL to form TEABT ↗

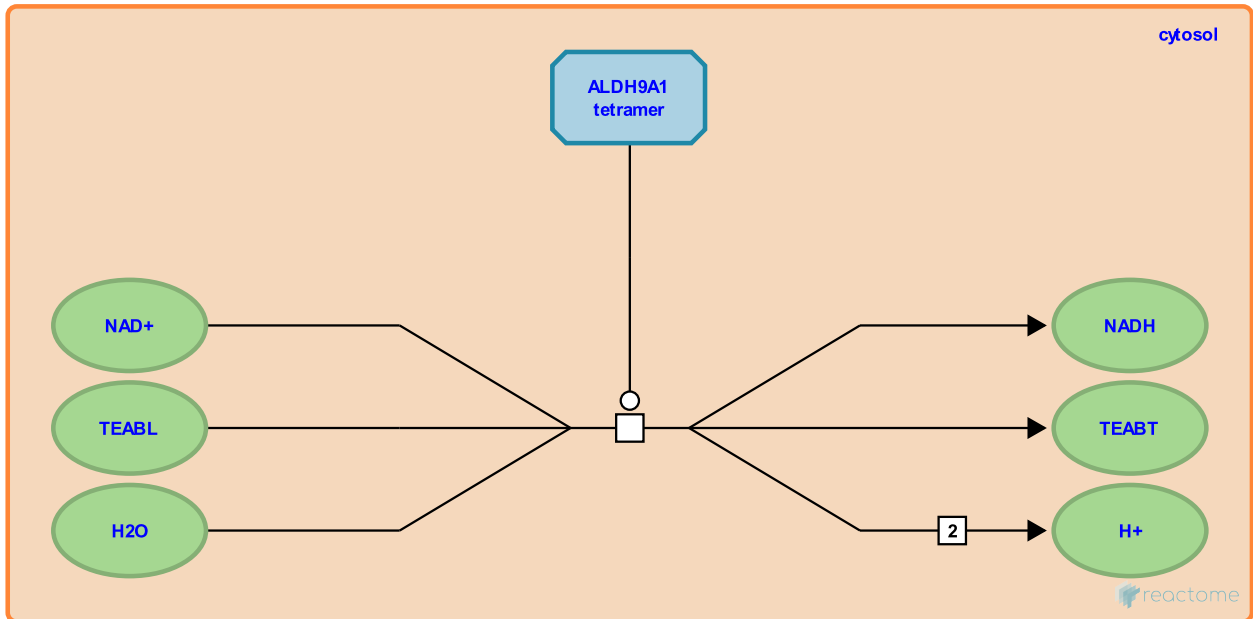
Location: [Carnitine synthesis](#)

Stable identifier: R-CFA-71260

Type: transition

Compartments: cytosol

Inferred from: [ALDH9A1 tetramer dehydrogenates TEABL to form TEABT \(Homo sapiens\)](#)



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: [SHMT1 tetramer cleaves HTMLYS to yield TEABL and Gly](#)

Followed by: [BBOX1:AscH-:Fe2+ dimer dioxygenates TEABT and 2OG to form CAR and SUCCA](#)

BBOX1:AscH-:Fe2+ dimer dioxygenates TEABT and 2OG to form CAR and SUCCA ↗

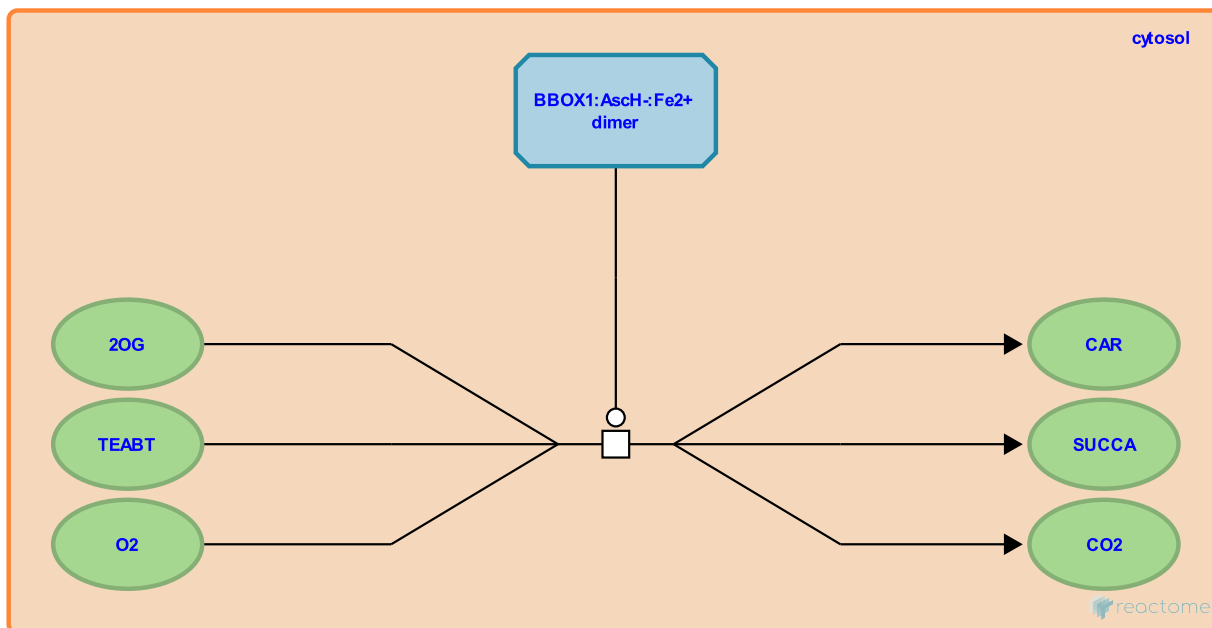
Location: [Carnitine synthesis](#)

Stable identifier: R-CFA-71261

Type: transition

Compartments: cytosol

Inferred from: [BBOX1:AscH-:Fe2+ dimer dioxygenates TEABT and 2OG to form CAR and SUCCA \(Homo sapiens\)](#)



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: [ALDH9A1 tetramer dehydrogenates TEABL to form TEABT](#)

Table of Contents

Introduction	1
☒ Carnitine synthesis	2
↳ TMLHE dimer dioxygenates TMLYS and 2OG to form HTMLYS and SUCCA	3
↳ SHMT1 tetramer cleaves HTMLYS to yield TEABL and Gly	4
↳ ALDH9A1 tetramer dehydrogenates TEABL to form TEABT	5
↳ BBOX1:AscH ⁻ :Fe ²⁺ dimer dioxygenates TEABT and 2OG to form CAR and SUCCA	6
Table of Contents	7