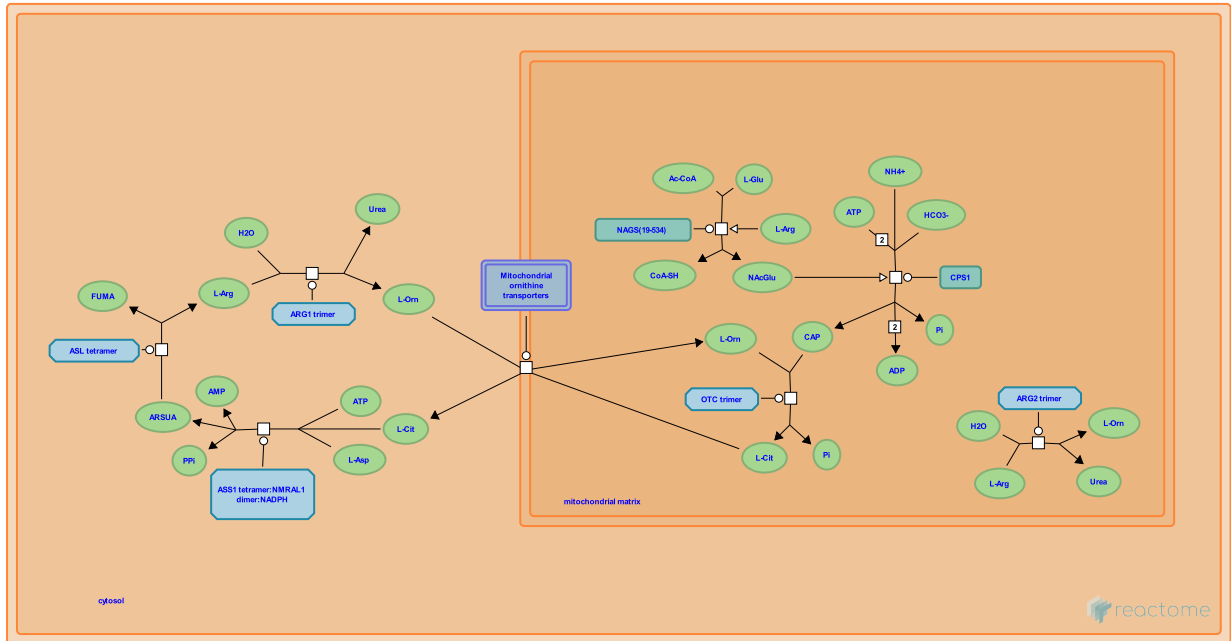


Urea cycle



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

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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/textbook/).

06/05/2024

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

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- 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. [↗](#)

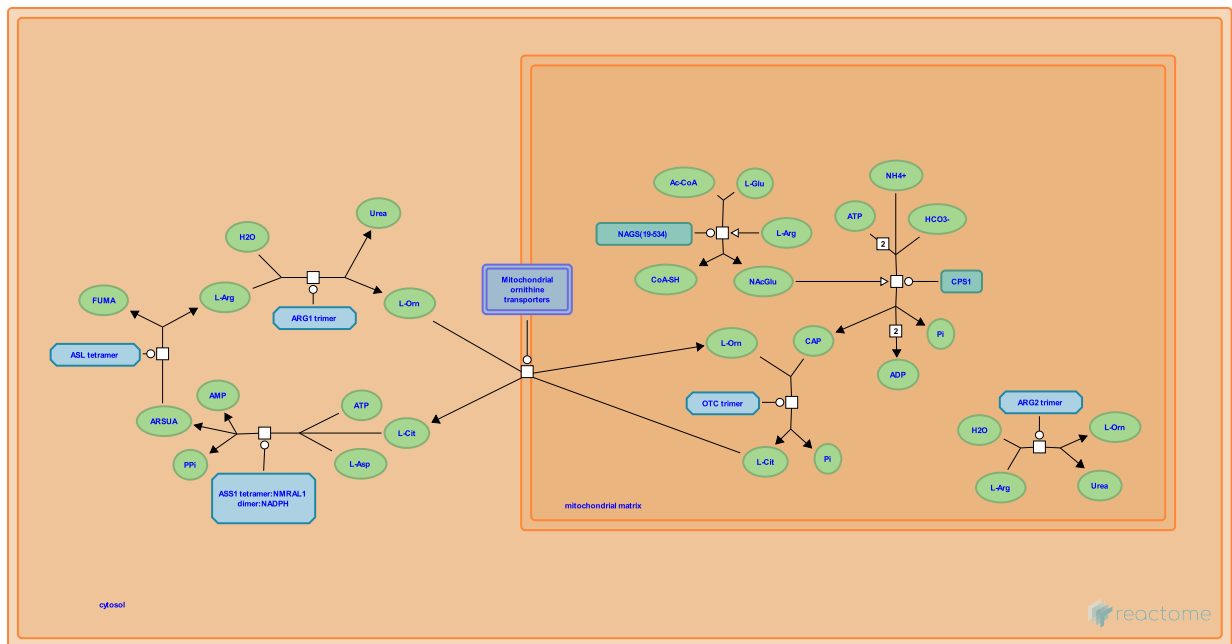
Reactome database release: 88

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

Urea cycle ↗

Stable identifier: R-BTA-70635

Inferred from: Urea cycle (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>

glutamate + acetyl CoA => N-acetyl glutamate + CoA ↗

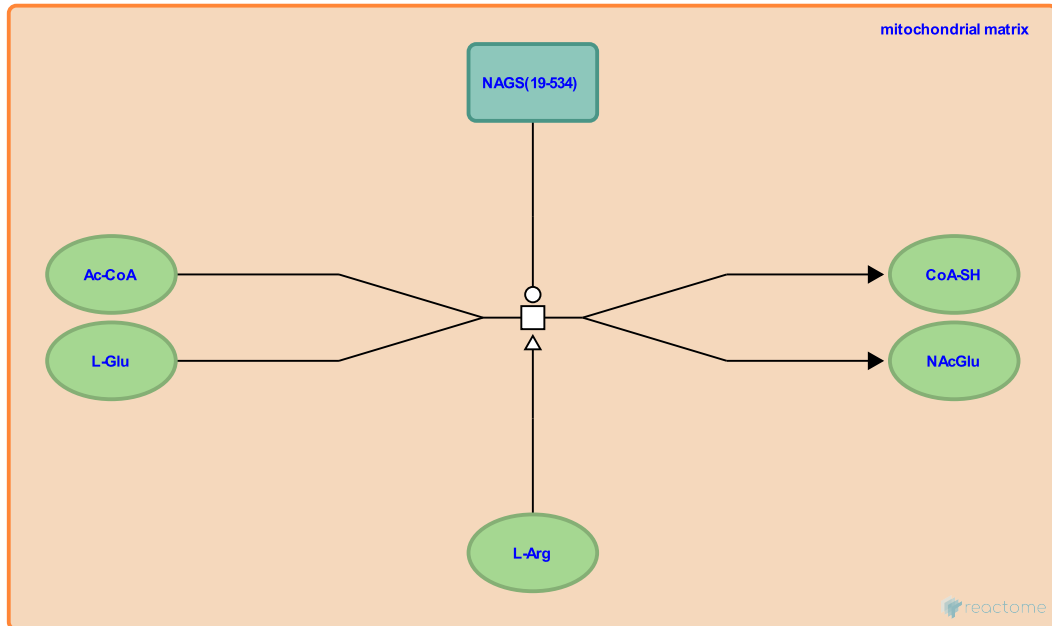
Location: Urea cycle

Stable identifier: R-BTA-70542

Type: transition

Compartments: mitochondrial matrix

Inferred from: glutamate + acetyl CoA => N-acetyl glutamate + 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>

Followed by: 2 ATP + NH₄⁺ + HCO₃⁻ => 2 ADP + orthophosphate + carbamoyl phosphate [mitochondrial]

2 ATP + NH₄⁺ + HCO₃⁻ => 2 ADP + orthophosphate + carbamoyl phosphate [mitochondrial] ↗

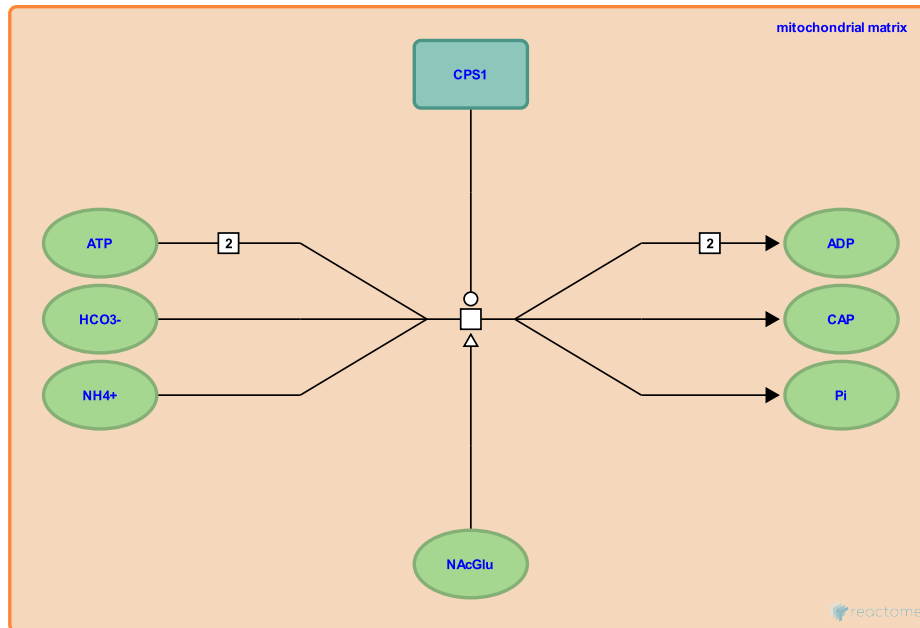
Location: Urea cycle

Stable identifier: R-BTA-70555

Type: transition

Compartments: mitochondrial matrix

Inferred from: 2 ATP + NH₄⁺ + HCO₃⁻ => 2 ADP + orthophosphate + carbamoyl phosphate [mitochondrial] (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: glutamate + acetyl CoA => N-acetyl glutamate + CoA

Followed by: carbamoyl phosphate + ornithine => citrulline + orthophosphate

carbamoyl phosphate + ornithine => citrulline + orthophosphate ↗

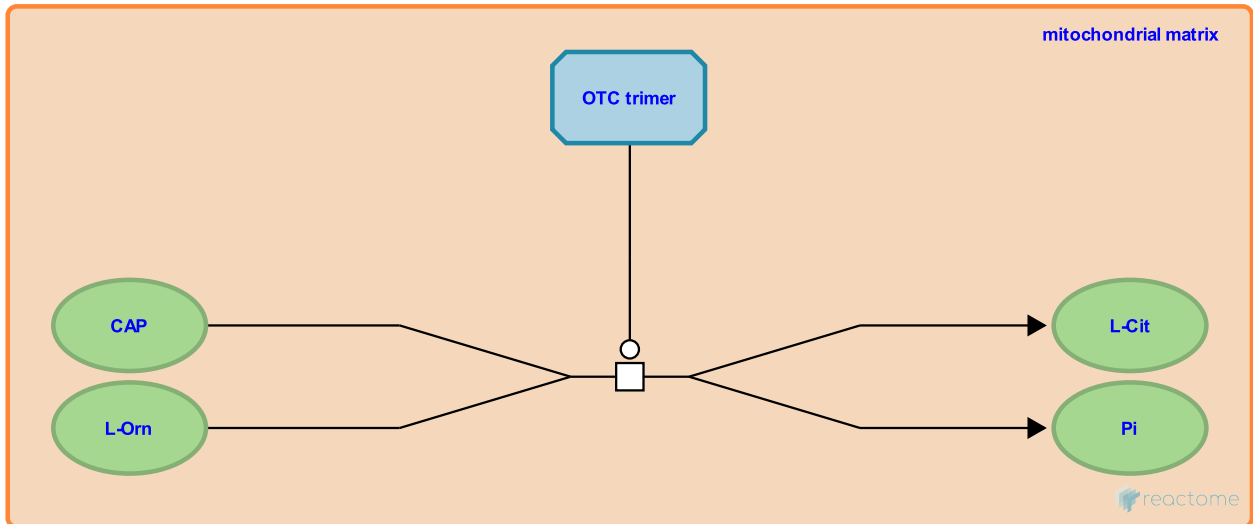
Location: Urea cycle

Stable identifier: R-BTA-70560

Type: transition

Compartments: mitochondrial matrix

Inferred from: carbamoyl phosphate + ornithine => citrulline + orthophosphate (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: ornithine (cytosolic) + citrulline (mitochondrial) => ornithine (mitochondrial) + citrulline (cytosolic), 2 ATP + NH₄⁺ + HCO₃⁻ => 2 ADP + orthophosphate + carbamoyl phosphate [mitochondrial]

Followed by: ornithine (cytosolic) + citrulline (mitochondrial) => ornithine (mitochondrial) + citrulline (cytosolic)

ASS1 tetramer:NMRAL1 dimer:NADPH transforms L-Asp and L-Cit to ARSUA ↗

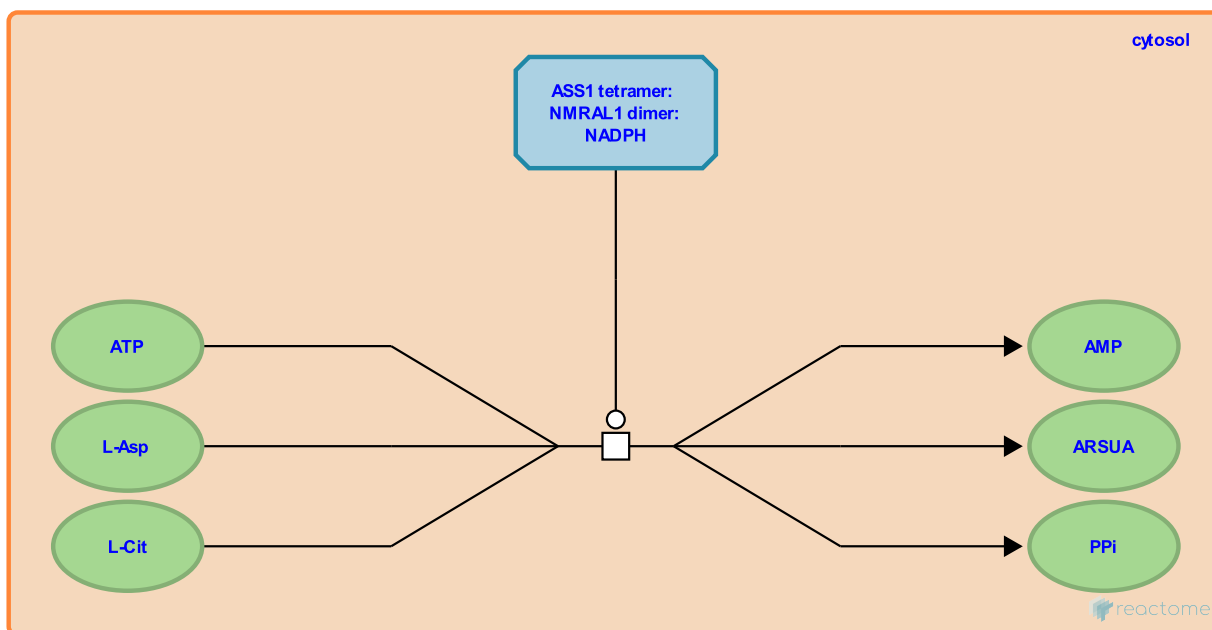
Location: Urea cycle

Stable identifier: R-BTA-70577

Type: transition

Compartments: cytosol

Inferred from: ASS1 tetramer:NMRAL1 dimer:NADPH transforms L-Asp and L-Cit to ARSUA (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: ornithine (cytosolic) + citrulline (mitochondrial) => ornithine (mitochondrial) + citrulline (cytosolic)

Followed by: argininosuccinate <=> fumarate + arginine

argininosuccinate <=> fumarate + arginine ↗

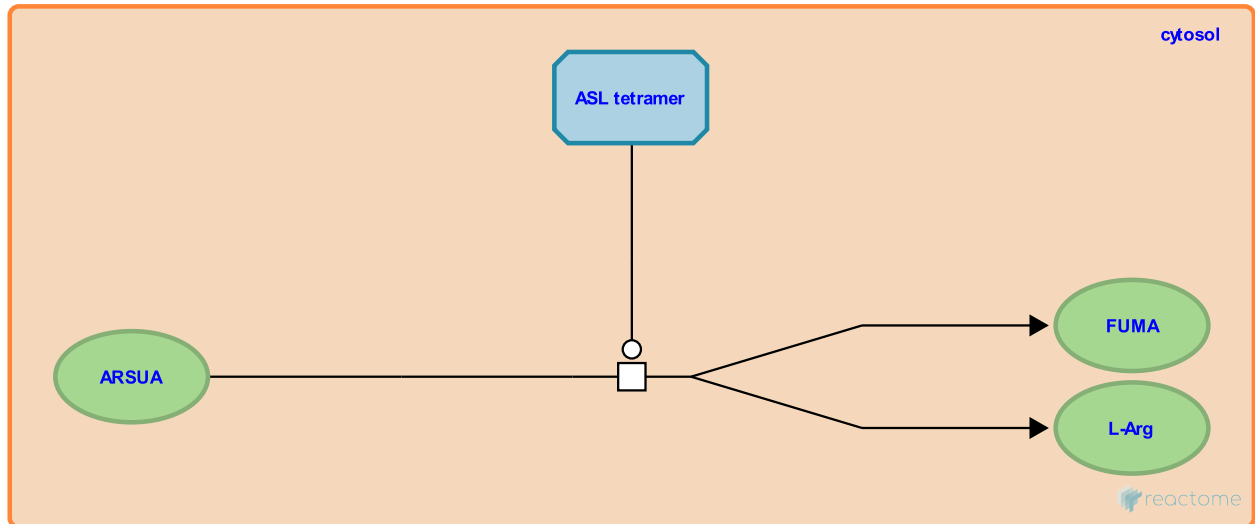
Location: Urea cycle

Stable identifier: R-BTA-70573

Type: transition

Compartments: cytosol

Inferred from: argininosuccinate <=> fumarate + arginine (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: ASS1 tetramer:NMRAL1 dimer:NADPH transforms L-Asp and L-Cit to ARSUA

Followed by: arginine + H₂O => ornithine + urea [ARG1]

arginine + H₂O => ornithine + urea [ARG1] ↗

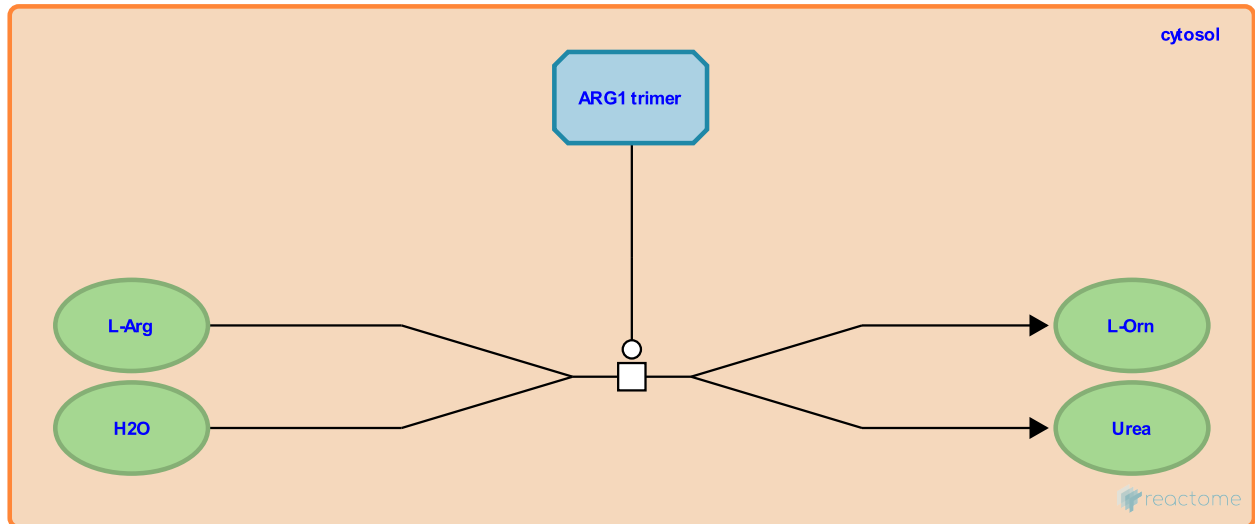
Location: Urea cycle

Stable identifier: R-BTA-70569

Type: transition

Compartments: cytosol

Inferred from: arginine + H₂O => ornithine + urea [ARG1] (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: argininosuccinate <=> fumarate + arginine

Followed by: ornithine (cytosolic) + citrulline (mitochondrial) => ornithine (mitochondrial) + citrulline (cytosolic)

arginine + H₂O => ornithine + urea [ARG2] ↗

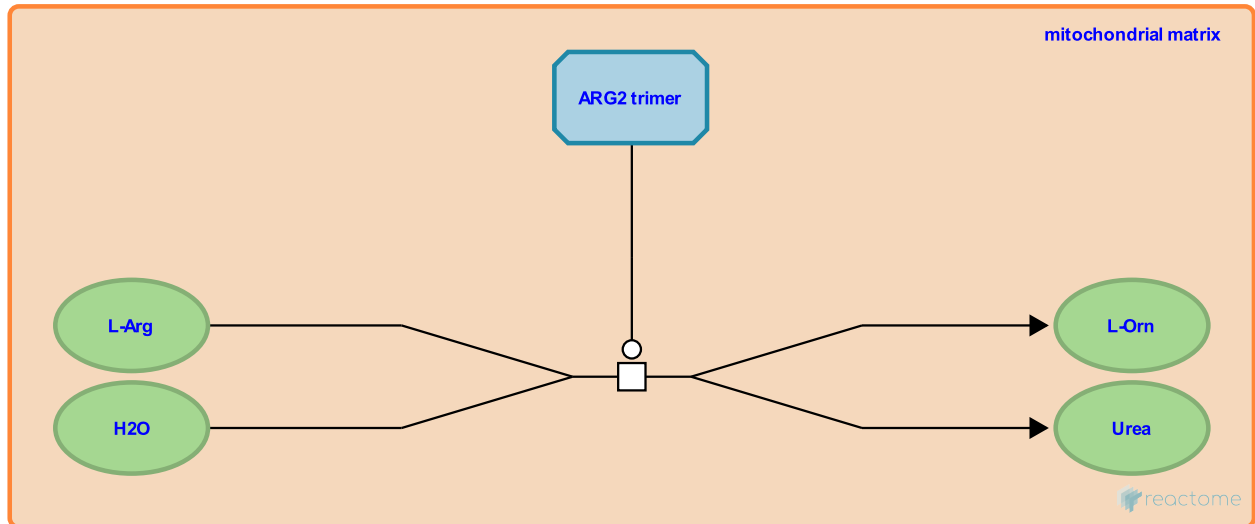
Location: Urea cycle

Stable identifier: R-BTA-452036

Type: transition

Compartments: mitochondrial matrix

Inferred from: arginine + H₂O => ornithine + urea [ARG2] (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>

ornithine (cytosolic) + citrulline (mitochondrial) => ornithine (mitochondrial) + citrulline (cytosolic) ↗

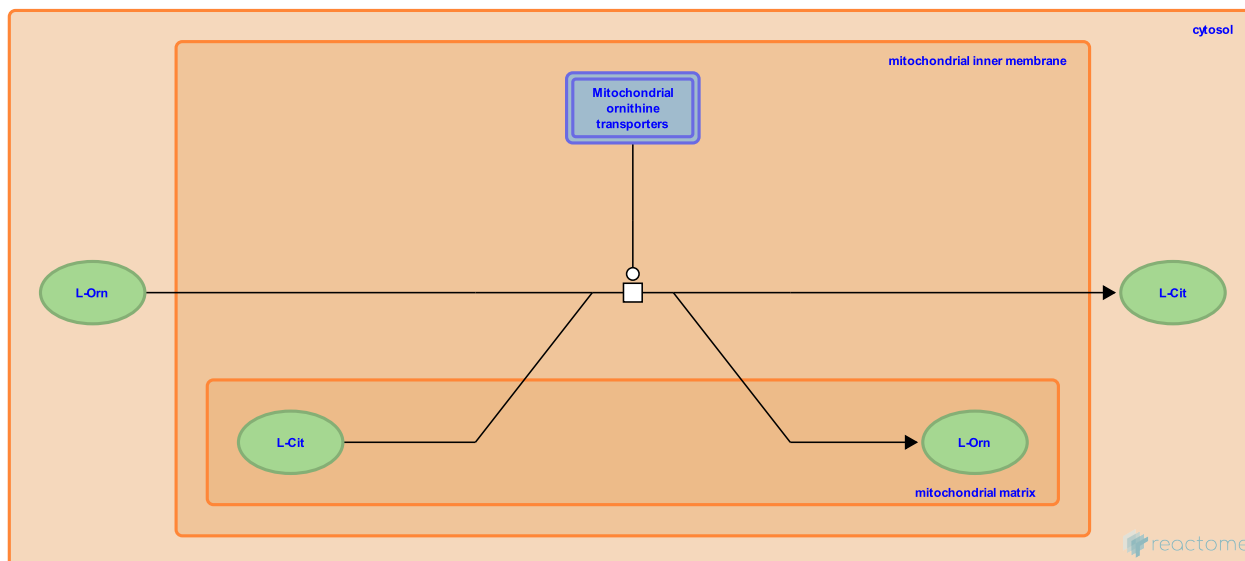
Location: Urea cycle

Stable identifier: R-BTA-70634

Type: transition

Compartments: mitochondrial inner membrane, mitochondrial matrix, cytosol

Inferred from: ornithine (cytosolic) + citrulline (mitochondrial) => ornithine (mitochondrial) + citrulline (cytosolic) (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: arginine + H₂O => ornithine + urea [ARG1], carbamoyl phosphate + ornithine => citrulline + orthophosphate

Followed by: ASS1 tetramer:NMRAL1 dimer:NADPH transforms L-Asp and L-Cit to ARSUA, carbamoyl phosphate + ornithine => citrulline + orthophosphate

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