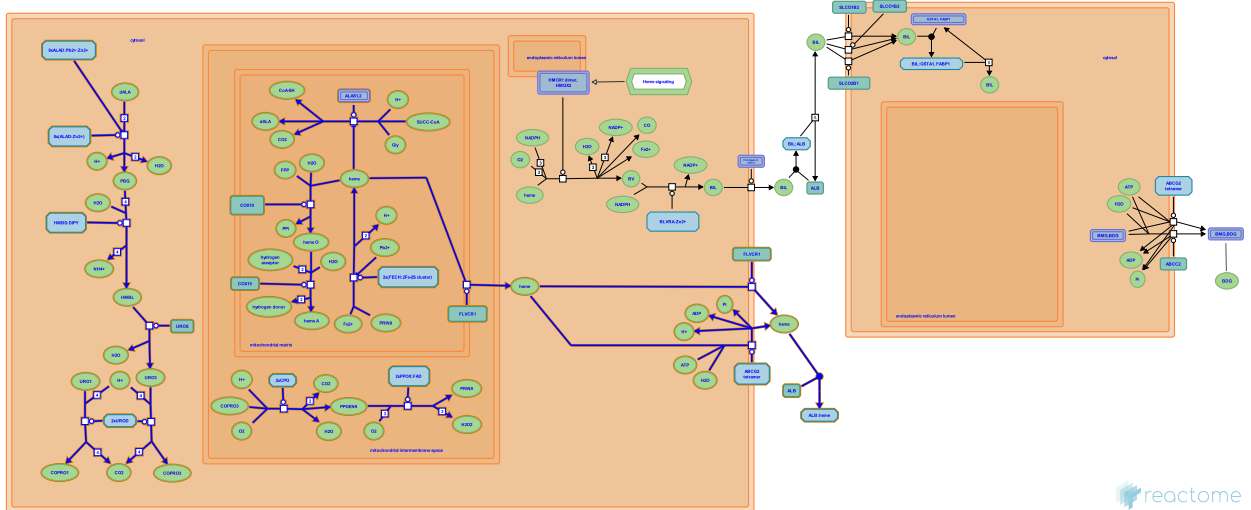


# Heme biosynthesis



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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/).

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

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- Sidiropoulos, K., Viteri, G., Sevilla, C., Jupe, S., Webber, M., Orlic-Milacic, M. et al. (2017). Reactome enhanced pathway visualization. *Bioinformatics*, 33, 3461-3467. [↗](#)
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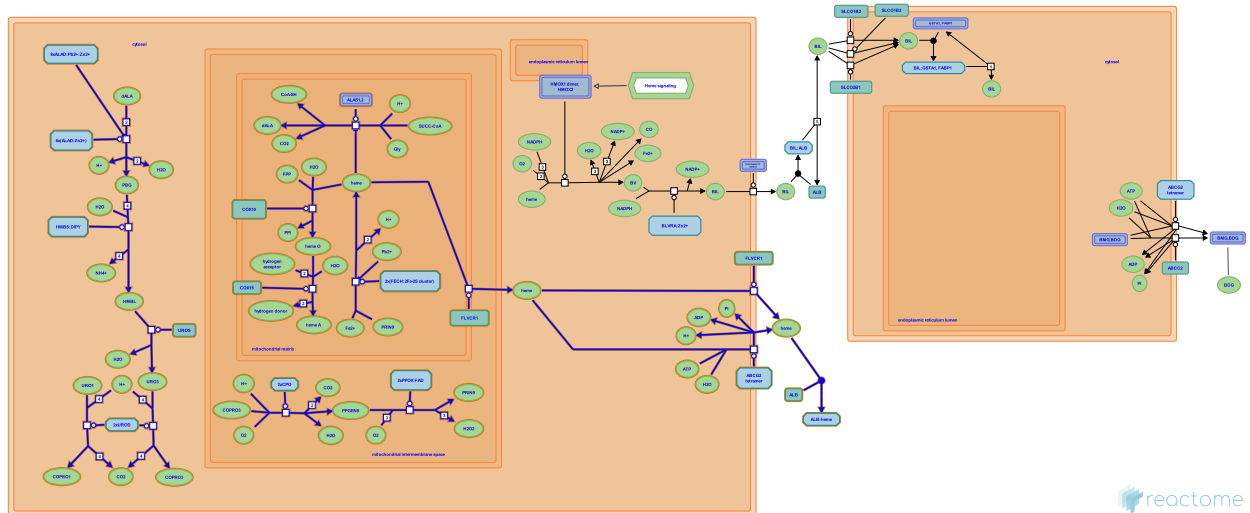
Reactome database release: 88

This document contains 1 pathway and 15 reactions ([see Table of Contents](#))

## Heme biosynthesis ↗

**Stable identifier:** R-CFA-189451

**Inferred from:** [Heme biosynthesis \(Homo sapiens\)](#)



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>

## ALAS condenses SUCC-CoA and Gly to form dALA [↗](#)

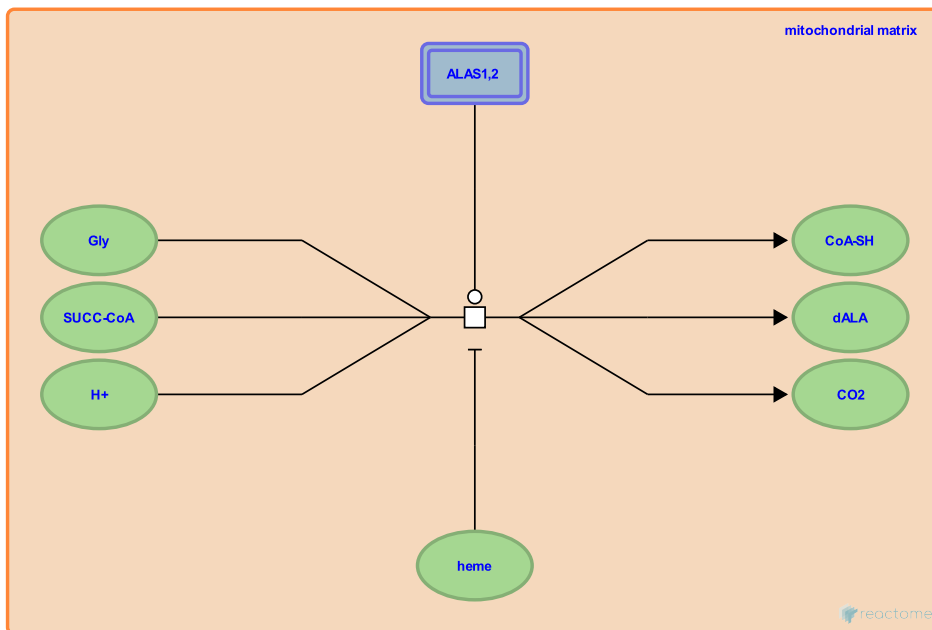
**Location:** [Heme biosynthesis](#)

**Stable identifier:** R-CFA-189442

**Type:** transition

**Compartments:** mitochondrial matrix

**Inferred from:** [ALAS condenses SUCC-CoA and Gly to form dALA \(Homo sapiens\)](#)



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>

## ALAD condenses 2 dALAs to form PBG ↗

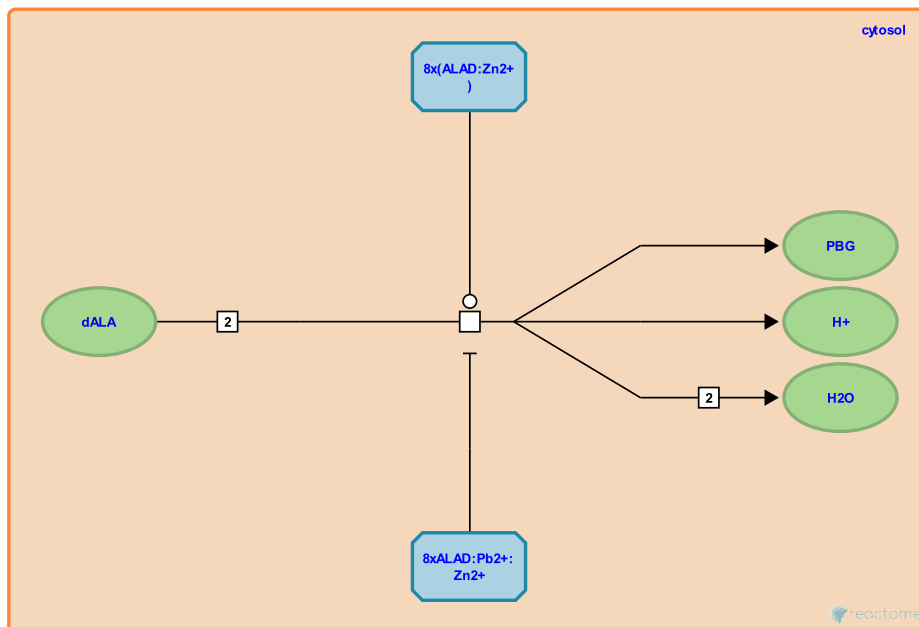
**Location:** [Heme biosynthesis](#)

**Stable identifier:** R-CFA-189439

**Type:** transition

**Compartments:** cytosol

**Inferred from:** [ALAD condenses 2 dALAs to form PBG \(Homo sapiens\)](#)



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:** [4 PBGs bind to form HMB](#)

#### 4 PBGs bind to form HMB ↗

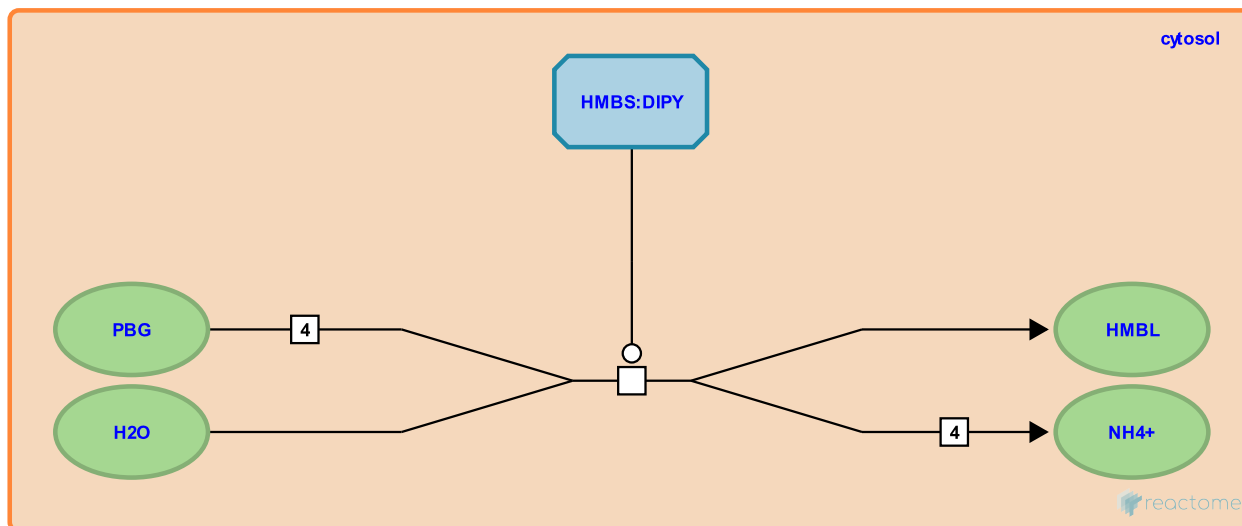
**Location:** [Heme biosynthesis](#)

**Stable identifier:** R-CFA-189406

**Type:** transition

**Compartments:** cytosol

**Inferred from:** [4 PBGs bind to form HMB \(Homo sapiens\)](#)



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:** [ALAD condenses 2 dALAs to form PBG](#)

**Followed by:** [UROS transforms HMB to URO3](#)

## UROS transforms HMB to URO3 ↗

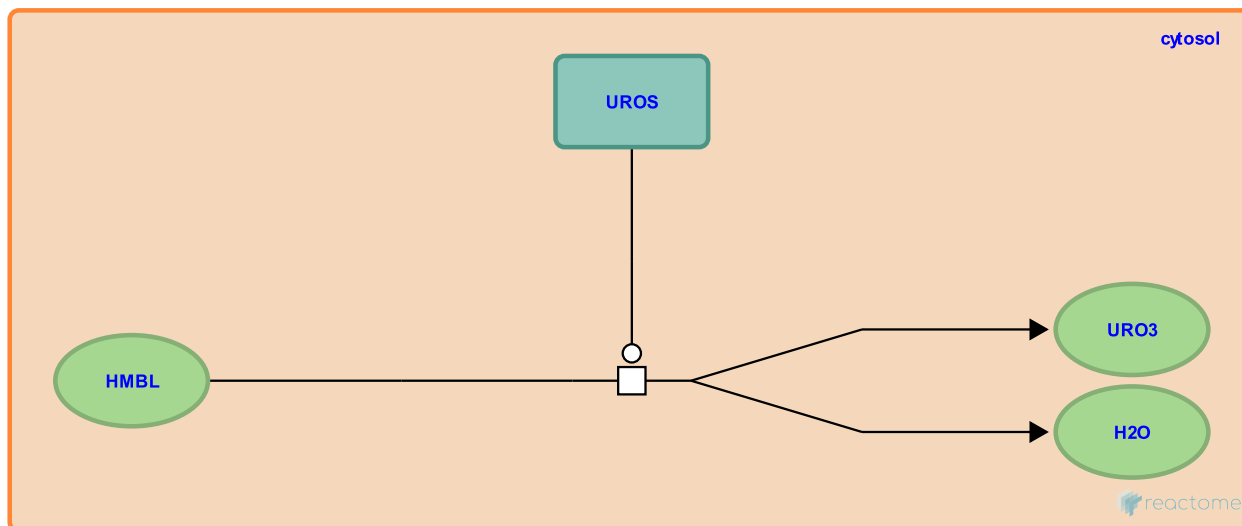
**Location:** [Heme biosynthesis](#)

**Stable identifier:** R-CFA-189488

**Type:** transition

**Compartments:** cytosol

**Inferred from:** [UROS transforms HMB to URO3 \(Homo sapiens\)](#)



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:** [4 PBGs bind to form HMB](#)

**Followed by:** [UROD decarboxylates URO3 to COPRO3](#)

## UROD decarboxylates URO3 to COPRO3 ↗

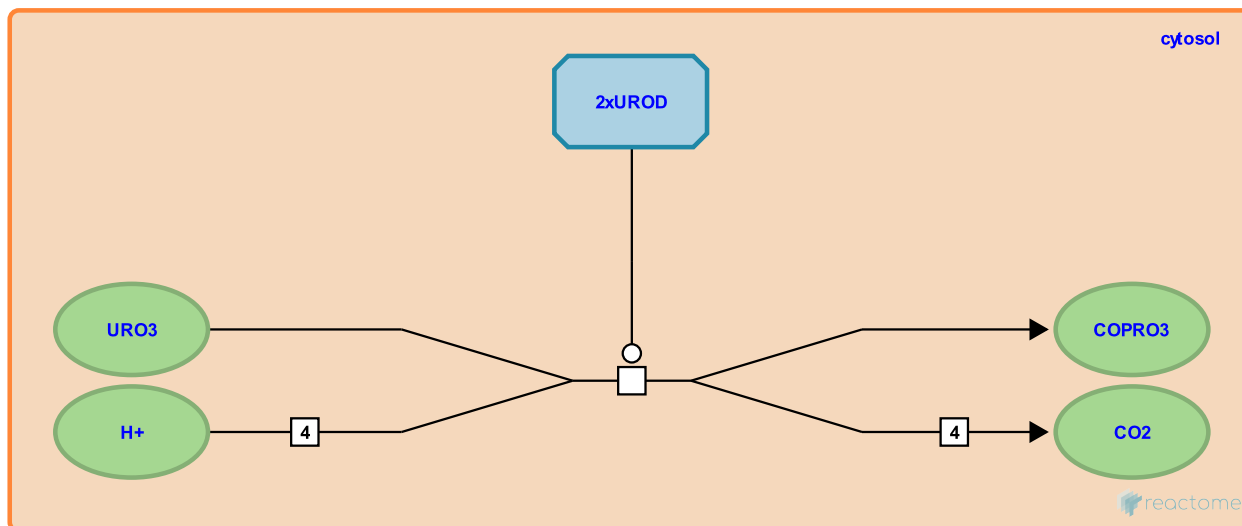
**Location:** [Heme biosynthesis](#)

**Stable identifier:** R-CFA-189425

**Type:** transition

**Compartments:** cytosol

**Inferred from:** [UROD decarboxylates URO3 to COPRO3 \(Homo sapiens\)](#)



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:** [UROS transforms HMB to URO3](#)



## UROD decarboxylates URO1 to COPRO1 ↗

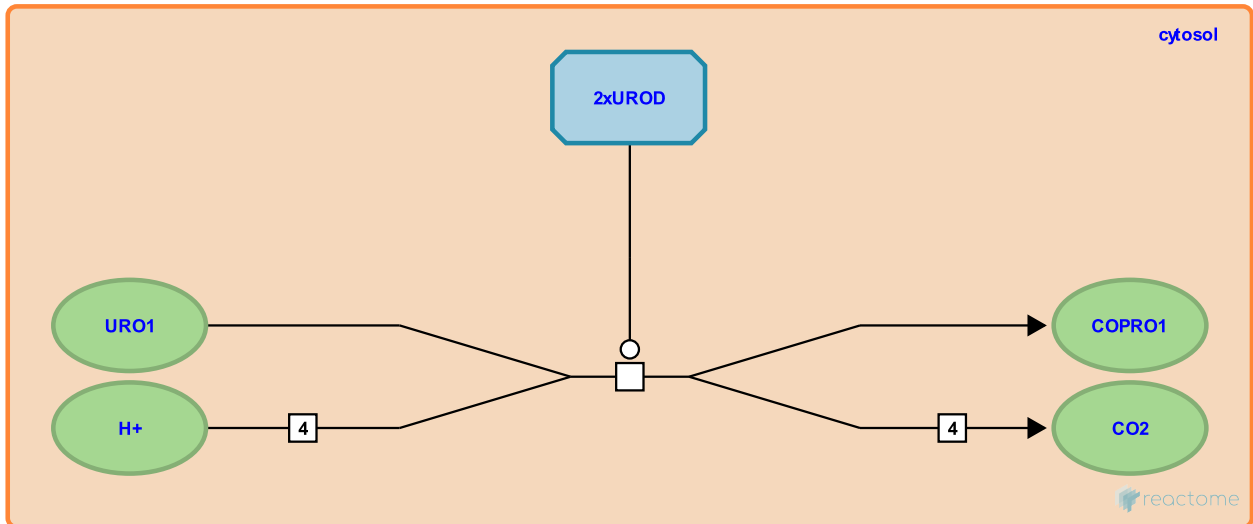
**Location:** [Heme biosynthesis](#)

**Stable identifier:** R-CFA-190182

**Type:** transition

**Compartments:** cytosol

**Inferred from:** [UROD decarboxylates URO1 to COPRO1 \(Homo sapiens\)](#)



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>

## CPO transforms COPRO3 to PPGEN9 ↗

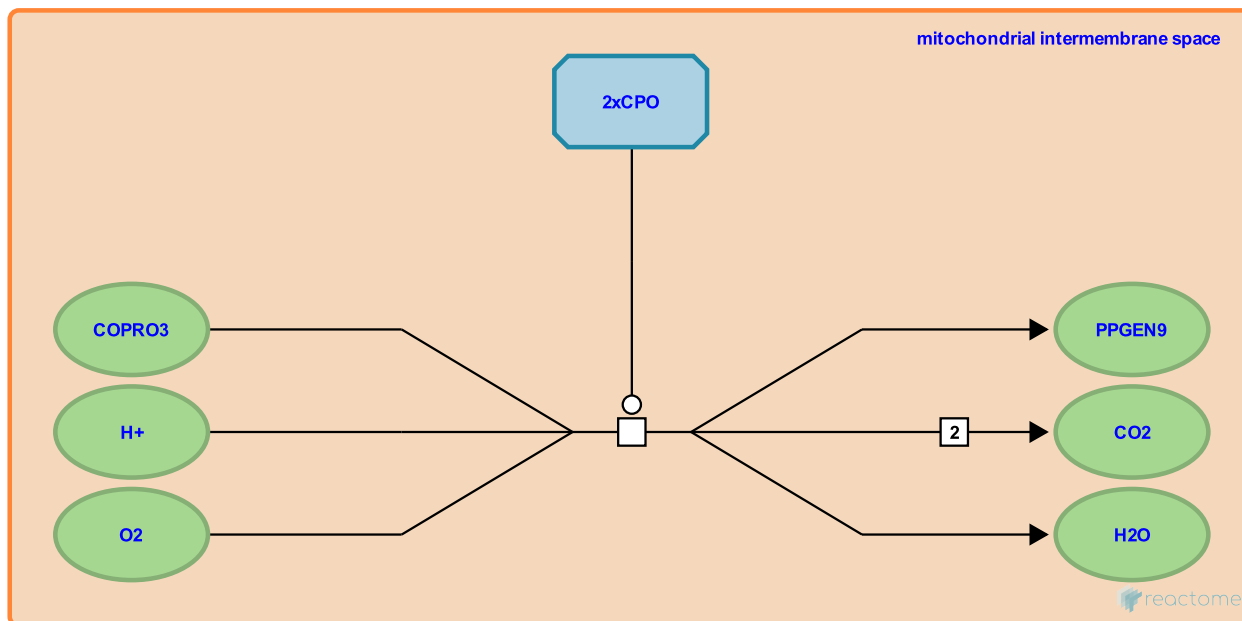
**Location:** [Heme biosynthesis](#)

**Stable identifier:** R-CFA-189421

**Type:** transition

**Compartments:** mitochondrial intermembrane space

**Inferred from:** [CPO transforms COPRO3 to PPGEN9 \(Homo sapiens\)](#)



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:** [PPO oxidises PPGEN9 to PRIN9](#)

## PPO oxidises PPGEN9 to PRIN9 ↗

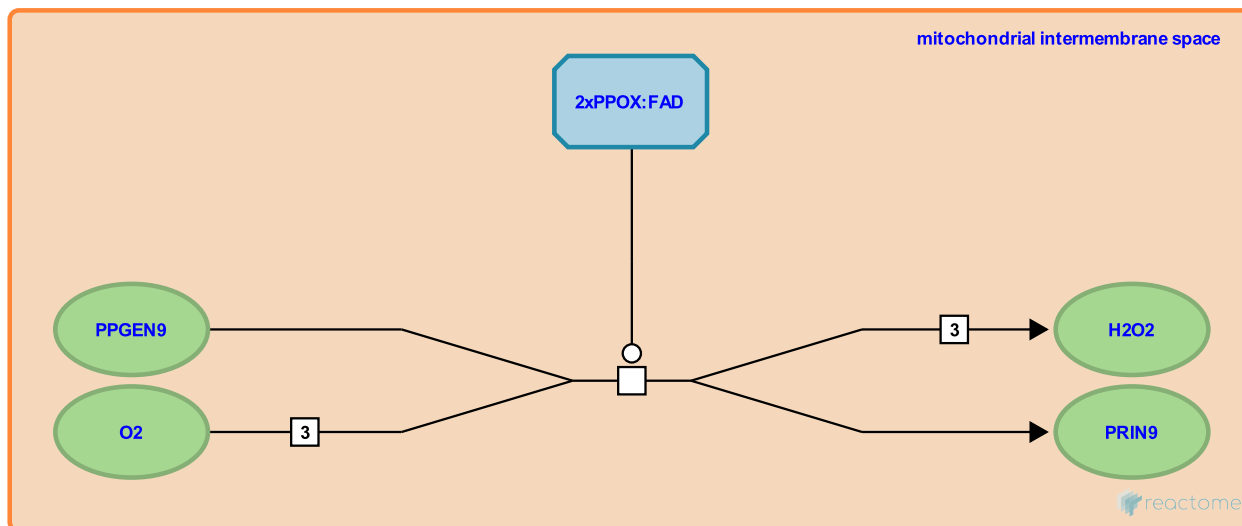
**Location:** [Heme biosynthesis](#)

**Stable identifier:** R-CFA-189423

**Type:** transition

**Compartments:** mitochondrial intermembrane space

**Inferred from:** [PPO oxidises PPGEN9 to PRIN9 \(Homo sapiens\)](#)



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:** [CPO transforms COPRO3 to PPGEN9](#)

## FECH binds Fe<sup>2+</sup> to PRIN9 to form heme ↗

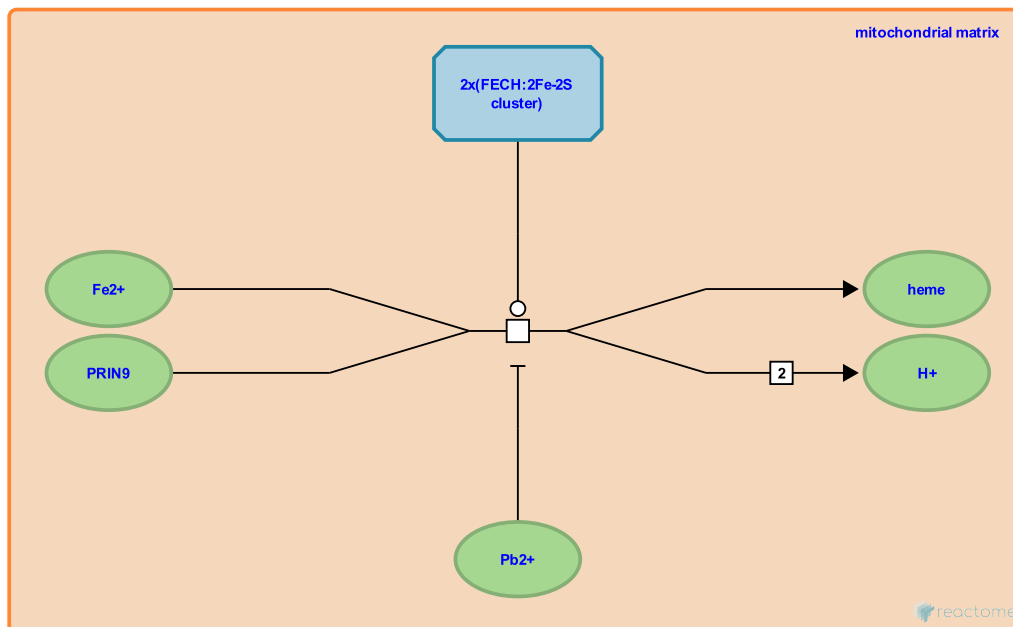
**Location:** [Heme biosynthesis](#)

**Stable identifier:** R-CFA-189465

**Type:** transition

**Compartments:** mitochondrial matrix

**Inferred from:** [FECH binds Fe<sup>2+</sup> to PRIN9 to form heme \(Homo sapiens\)](#)



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:** [COX10 transforms heme to heme O](#)

## FLVCR1-2 transports heme from mitochondrial matrix to cytosol ↗

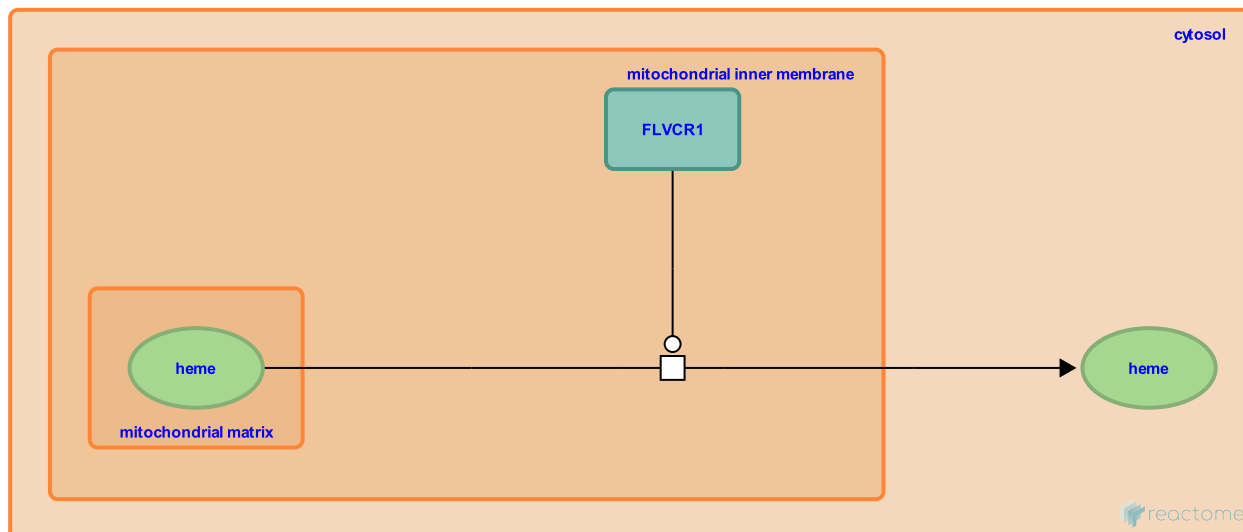
**Location:** [Heme biosynthesis](#)

**Stable identifier:** R-CFA-9661408

**Type:** transition

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

**Inferred from:** [FLVCR1-2 transports heme from mitochondrial matrix 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>

## ABCG2 tetramer transports heme from cytosol to extracellular region ↗

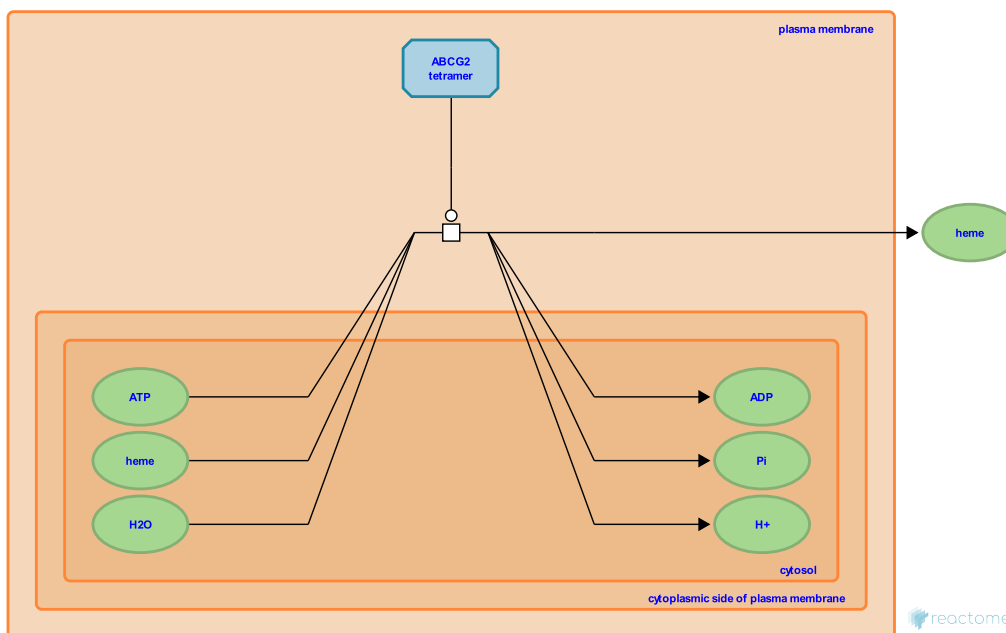
**Location:** [Heme biosynthesis](#)

**Stable identifier:** R-CFA-917979

**Type:** transition

**Compartments:** plasma membrane

**Inferred from:** [ABCG2 tetramer transports heme from cytosol to extracellular region \(Homo sapiens\)](#)



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>

## FLVCR1-1 transports heme from cytosol to extracellular region ↗

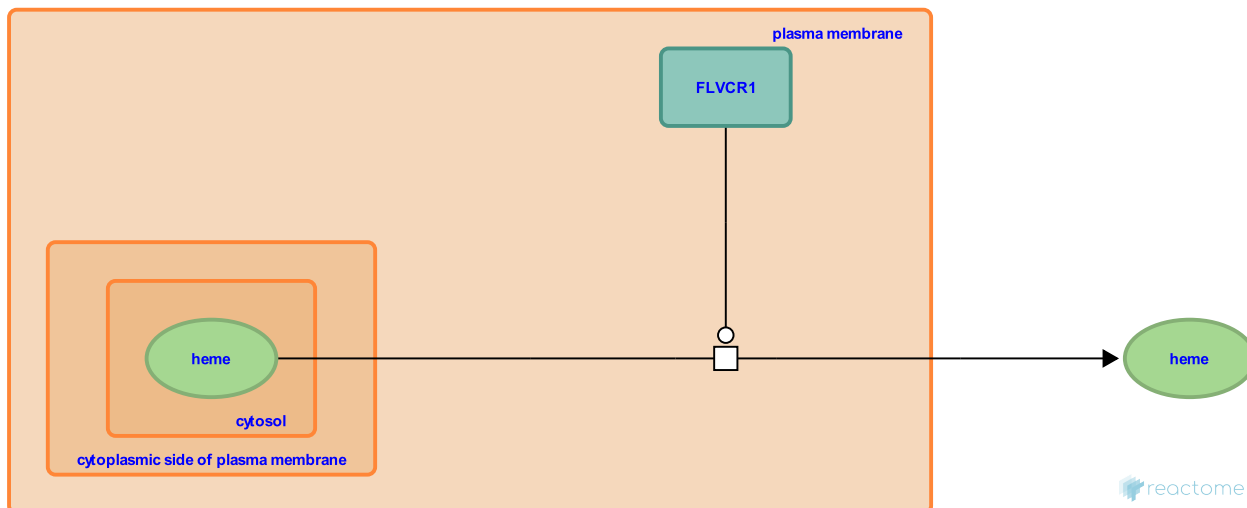
**Location:** [Heme biosynthesis](#)

**Stable identifier:** R-CFA-917892

**Type:** transition

**Compartments:** plasma membrane

**Inferred from:** [FLVCR1-1 transports heme from cytosol to extracellular region \(Homo sapiens\)](#)



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>

## ALB binds extracellular heme ↗

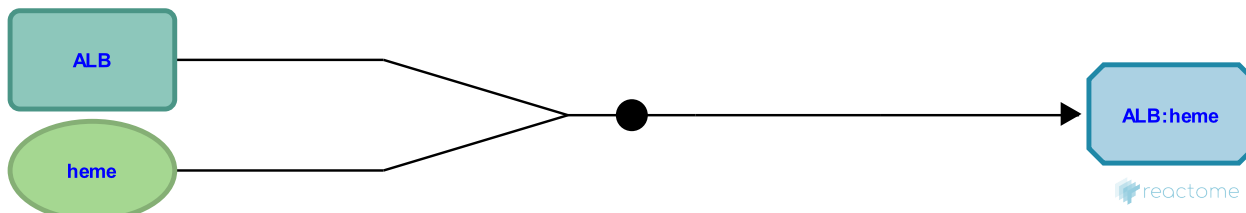
**Location:** [Heme biosynthesis](#)

**Stable identifier:** R-CFA-9661419

**Type:** binding

**Compartments:** extracellular region

**Inferred from:** [ALB binds extracellular heme \(Homo sapiens\)](#)



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>



## COX10 transforms heme to heme O [↗](#)

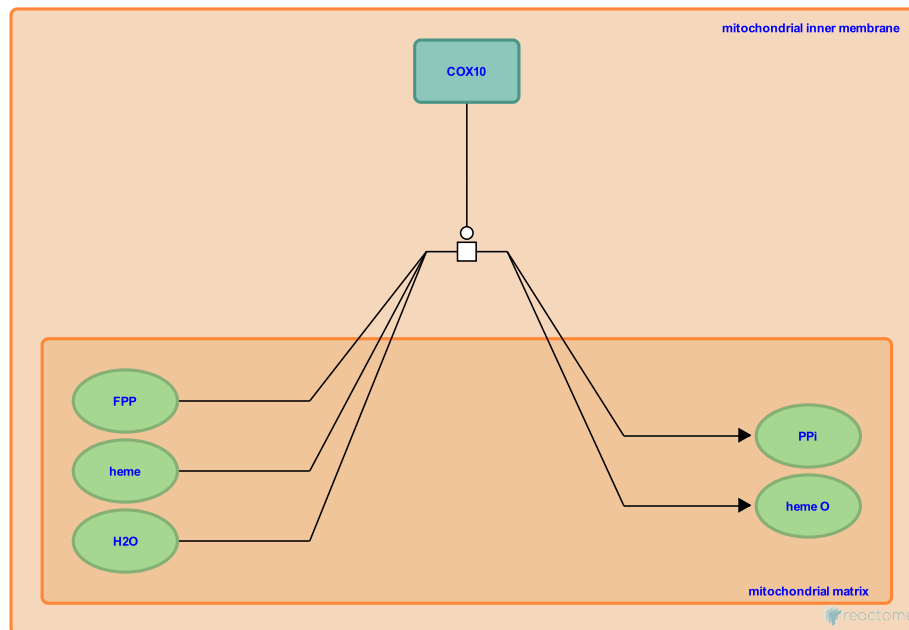
**Location:** [Heme biosynthesis](#)

**Stable identifier:** R-CFA-2995330

**Type:** transition

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

**Inferred from:** [COX10 transforms heme to heme O \(Homo sapiens\)](#)



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:** [FECH binds Fe<sup>2+</sup> to PRIN9 to form heme](#)

**Followed by:** [COX15 transforms heme O to heme A](#)

## COX15 transforms heme O to heme A ↗

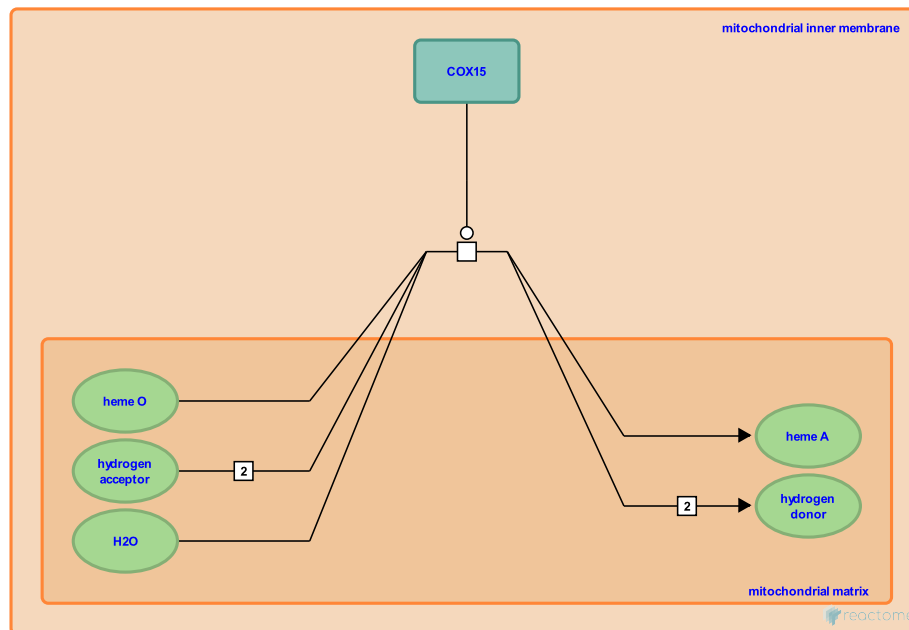
**Location:** [Heme biosynthesis](#)

**Stable identifier:** R-CFA-2995334

**Type:** transition

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

**Inferred from:** [COX15 transforms heme O to heme A \(Homo sapiens\)](#)



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:** [COX10 transforms heme to heme O](#)

# Table of Contents

Introduction	1
☰ Heme biosynthesis	2
↳ ALAS condenses SUCC-CoA and Gly to form dALA	3
↳ ALAD condenses 2 dALAs to form PBG	4
↳ 4 PBGs bind to form HMB	5
↳ UROS transforms HMB to URO3	6
↳ UROD decarboxylates URO3 to COPRO3	7
↳ UROD decarboxylates URO1 to COPRO1	8
↳ CPO transforms COPRO3 to PPGEN9	9
↳ PPO oxidises PPGEN9 to PRIN9	10
↳ FECH binds Fe <sup>2+</sup> to PRIN9 to form heme	11
↳ FLVCR1-2 transports heme from mitochondrial matrix to cytosol	12
↳ ABCG2 tetramer transports heme from cytosol to extracellular region	13
↳ FLVCR1-1 transports heme from cytosol to extracellular region	14
↳ ALB binds extracellular heme	15
↳ COX10 transforms heme to heme O	16
↳ COX15 transforms heme O to heme A	17
Table of Contents	18