

CPO transforms COPRO3 to PPGEN9

D'Eustachio, P., Hill, DP., Jassal, B., Sassa, S.

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 database: Efficient access to complex pathway data. *PLoS computational biology*, 14, e1005968. [↗](#)

Reactome database release: 88

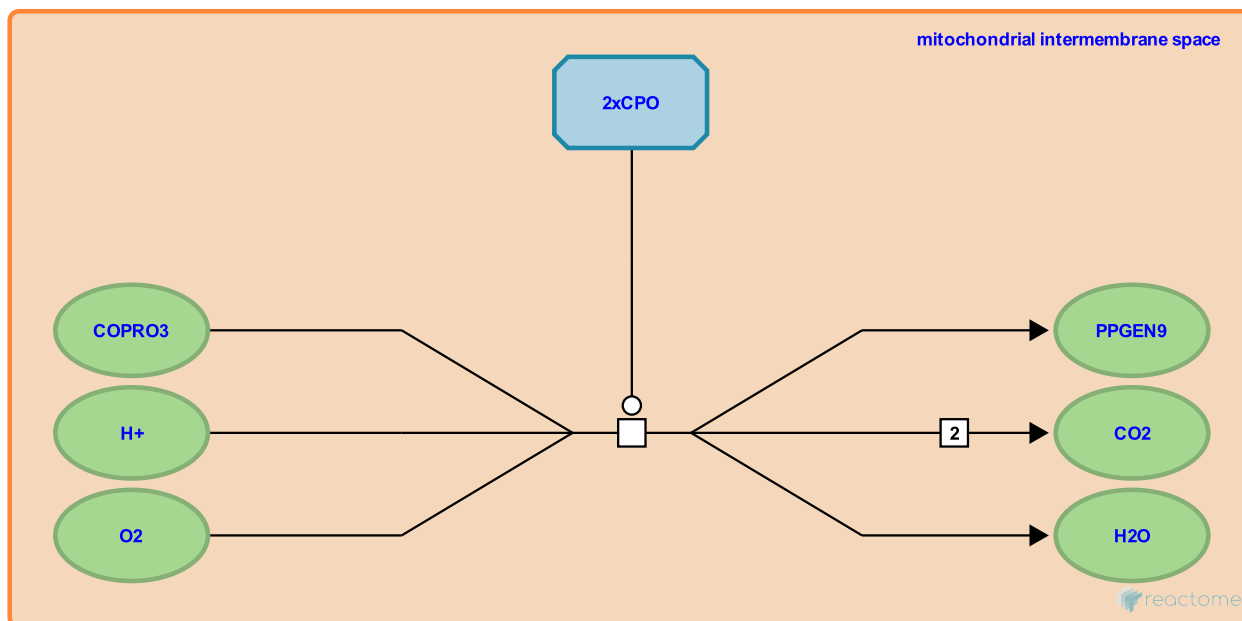
This document contains 1 reaction ([see Table of Contents](#))

CPO transforms COPRO3 to PPGEN9 [↗](#)

Stable identifier: R-HSA-189421

Type: transition

Compartments: mitochondrial intermembrane space



O₂-dependent coproporphyrinogen oxidase (CPO) catalyzes the conversion of coproporphyrinogen III (COPRO3) to protoporphyrinogen IX (PPGEN9) (Martasek et al. 1994). The localization of the human enzyme to the mitochondrial intermembrane space is inferred from studies of the homologous rat enzyme (Elder and Evans 1978). The human enzyme functions as a homodimer (Lee et al. 2005). Enzyme deficiency is associated with hereditary coproporphyrinemia in vivo.

Literature references

- Grandchamp, B., Labbe, P., Dumas, JB., de Verneuil, H., Delfau-Larue, MH., Montagne, JJ. et al. (1994). Molecular cloning, sequencing, and functional expression of a cDNA encoding human coproporphyrinogen oxidase. *Proc Natl Acad Sci U S A*, 91, 3024-8. [↗](#)
- Evans, JO., Elder, GH. (1978). Evidence that the coproporphyrinogen oxidase activity of rat liver is situated in the intermembrane space of mitochondria. *Biochem J*, 172, 345-7. [↗](#)
- Cooper, CL., Jones, MA., Lash, TD. (2005). Kinetic evaluation of human cloned coproporphyrinogen oxidase using a ring isomer of the natural substrate. *Med Sci Monit*, 11, BR420-5. [↗](#)
- Lee, DS., Martasek, P., Flachsova, E., Demeler, B., Bodnarova, M., Raman, CS. (2005). Structural basis of hereditary coproporphyrinemia. *Proc Natl Acad Sci U S A*, 102, 14232-7. [↗](#)

Editions

2007-01-24	Authored, Edited	Jassal, B., D'Eustachio, P.
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2009-05-19	Revised	D'Eustachio, P.
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