

# PPO oxidises PPGEN9 to PRIN9

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18/05/2024

## 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. [↗](#)

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Reactome database release: 88

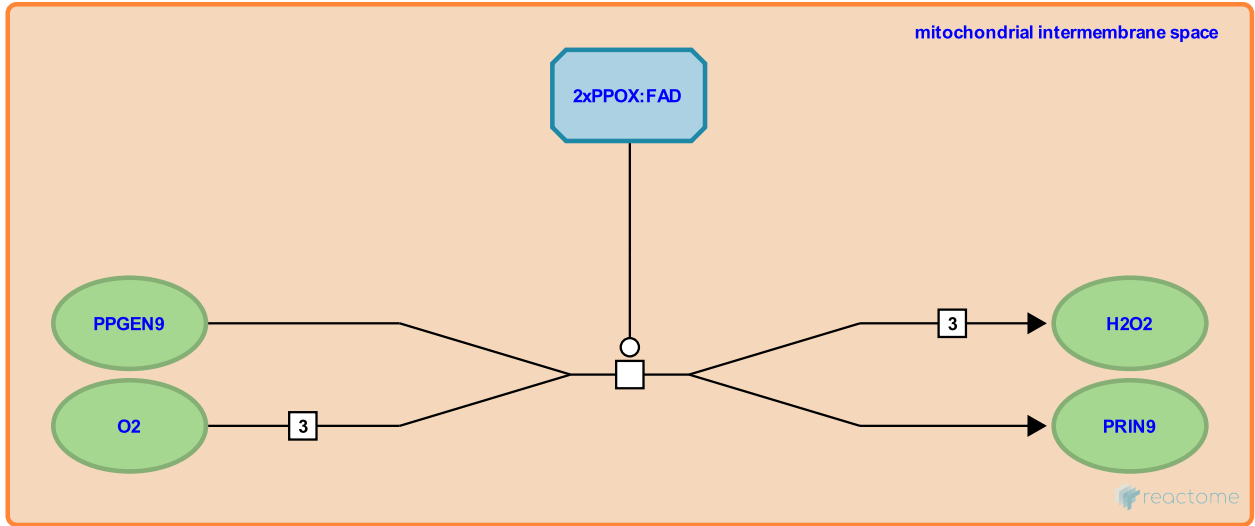
This document contains 1 reaction ([see Table of Contents](#))

**PPO oxidises PPGEN9 to PRIN9** ↗

**Stable identifier:** R-HSA-189423

**Type:** transition

**Compartments:** mitochondrial intermembrane space



Six electrons are oxidized in protoporphyrinogen IX (PPGEN9) to form the planar macrocycle protoporphyrin IX (PRIN9). This reaction is performed by the enzyme protoporphyrinogen oxidase (PPO). PPO functions as a homodimer containing one noncovalently-bound FAD. The protein resides on the outer surface of the inner mitochondrial membrane. PPO deficiency is associated with variegate porphyria in vivo (Dailey & Dailey 1996; Wang et al. 2013).

**Literature references**

Tan, Y., Wen, X., Wang, B., Qin, X., Shen, Y., Wang, Z. et al. (2013). Quantitative structural insight into human variegate porphyria disease. *J Biol Chem*, 288, 11731-40. ↗

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**Editions**

2007-01-24	Authored, Edited	Jassal, B., D'Eustachio, P.
2007-01-24	Reviewed	Sassa, S.
2009-05-19	Revised	D'Eustachio, P.
2023-11-22	Revised	D'Eustachio, P.