

PPO oxidises PPGEN9 to PRIN9

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

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/licenses/).

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

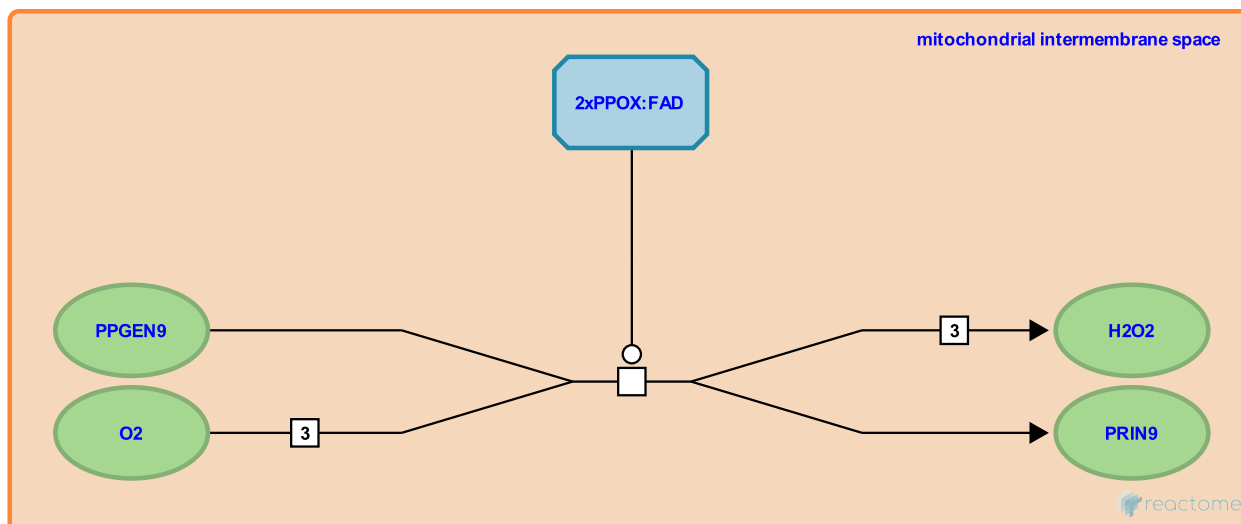
This document contains 1 reaction ([see Table of Contents](#))

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. ↗

Dailey, TA., Dailey, HA. (1996). Human protoporphyrinogen oxidase: expression, purification, and characterization of the cloned enzyme. *Protein Sci*, 5, 98-105. ↗

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.