4 PBGs bind to form HMB

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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

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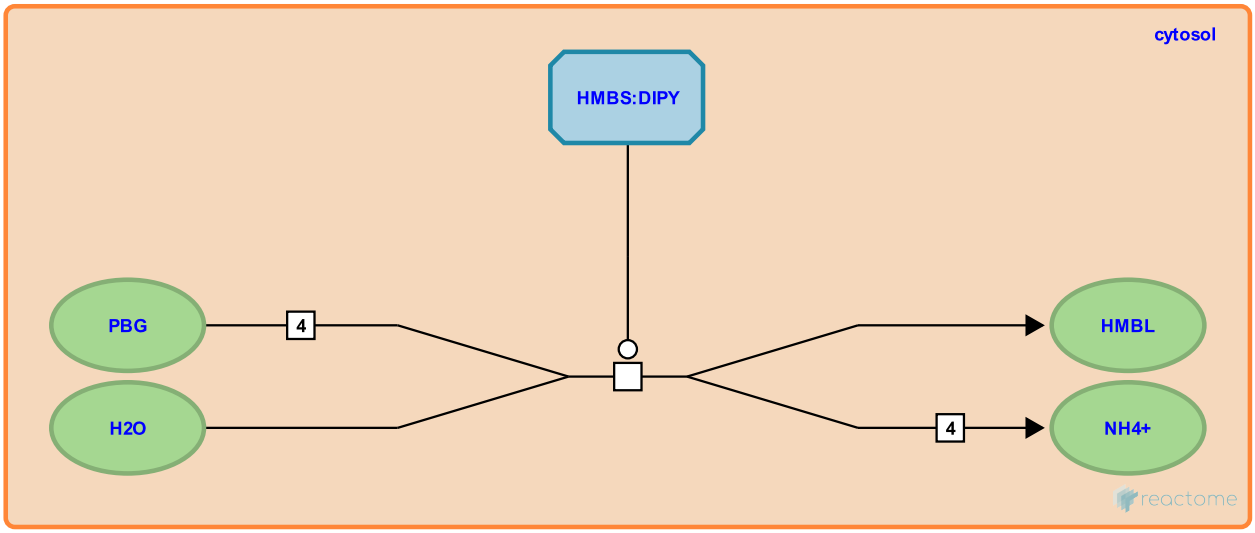
Stable identifier: R-HSA-189406

Type: transition

Compartments: cytosol



Cytosolic porphobilinogen deaminase catalyzes the polymerization of four molecules of porphobilinogen (PBG) to generate hydroxymethylbilane (HMB), an unstable tetrapyrrole. This reaction is the first step in the formation of the tetrapyrrole macrocycle. Two isoforms of porphobilinogen deaminase are generated by alternative splicing, one expresssed in erythroid tissues and one ubiquitously expressed in the body. Deficiencies of both forms of PBG deaminase are associated with acute intermittent porphyria.

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Editions

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