

# Porphobilinogen Deaminase

Human, PBGD

Expressed in *E. Coli*

Cat. No. CBCRY24

Lot. No. (See product label)

## BACKGROUND

The protein is the third enzyme of the heme biosynthetic pathway and catalyzes the head to tail condensation of four porphobilinogen molecules into the linear hydroxymethylbilane. Mutations in this gene are associated with the autosomal dominant disease acute intermittent porphyria. Alternatively spliced transcript variants encoding different isoforms have been described.

## MOLECULAR DESCRIPTION

**Protein classification:** Transferase

**Structure Weight:** 80152.23 Da

**Polymer:** 1

**Molecule:** Porphobilinogen deaminase

**Chains:** A, B

**Type:** polypeptide (L)

**Chain Length:** 364 amino acids

## CRYSTAL INFORMATION

**PDB ID:** [3ECR](#)

**MMDB ID:** [67075](#)

**Source:** E. Coli

**Method:** X-Ray Diffraction

**Resolution:** 2.18 Å

**Ligand Chemical Component:** DPM

## PRIMARY CITATION

Song, G., Li, Y., Cheng, C., Zhao, Y., Gao, A., Zhang, R., Joachimiak, A., Shaw, N., Liu, Z.J. (2009) Structural insight into acute intermittent porphyria. *Faseb J.* 23: 396-404

## FOR RESEARCH USE ONLY

## CRYSTAL STRUCTURE



## GENE INFORMATION

**Gene Name:** [HMBS](#)

**Synonyms:** PBG-D; PBGD; UPS; EC 2.5.1.61; porphobilinogen deaminase; pre-uroporphyrinogen synthase; uroporphyrinogen I synthase; uroporphyrinogen I synthetase; hydroxymethylbilane synthase

**UniProt ID:** [P08397](#)

**GeneID:** [3145](#)

**Chromosome Location:** 11q23.3

**Function:** hydroxymethylbilane synthase activity; transferase activity