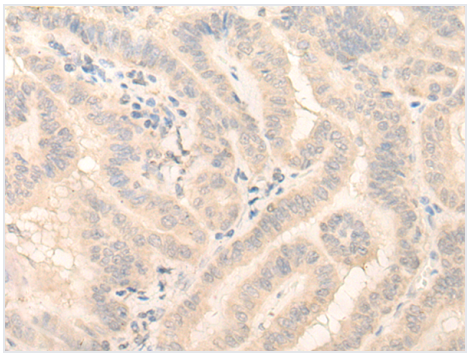
HMBS Antibody – Rabbit Polyclonal

PRODUCT OVERVIEW

Product Name	HMBS, Polyclonal Antibody
Full Product Name	HMBS Polyclonal Antibody
Product Gene Name	anti-HMBS antibody
Product Synonym Names	HEM3; HMBS; Hydroxymethylbilane synthase; PBG D; PBG-D; PBGD; PORC; Porphobilinogen deaminase; Porphyria; Acute; Chester type; Pre uroporphyrinogen synthase; Pre-uroporphyrinogen synthase; UPS; Uroporphyrinogen I synthase; Uroporphyrinogen I synthetase
Clonality	Polyclonal
Isotype	IgG
Host	Rabbit
Species Reactivity	Human, Mouse, Rat
Form/Format	PBS with 0.05% NaN3 and 40% Glycerol, pH7.4
Purity/Purification	Antigen Affinity Purification
Concentration	0.78mg/mL (lot specific)
Preparation and Storage	Store at -20 degree C. Avoid freeze/thaw cycles.
OMIM	phenotype 609806
3D Structure	ModBase 3D Structure for P08397
Immunogen	Fusion protein of human HMBS
Conjugation	Unconjugated
Product Categories/Family for anti-HMBS antibody	Cancer; Cell Biology; Metabolism; Signal transduction
ISO Certification	Manufactured in an ISO 9001:2015 Certified Laboratory.
Applications Tested/Suitable for anti-HMBS antibody	Immunohistochemistry (IHC), ELISA (EIA)
Application Notes for anti-HMBS antibody	IHC: 1:30-1:150 ELISA: 1:5000-1:10000
Related Product Information for anti-HMBS antibody	This gene encodes a member of the hydroxymethylbilane synthase superfamily. The encoded 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.
Other Notes	Small volumes of anti-HMBS antibody vial(s) may occasionally become entrapped in the seal of the product vial during shipment and storage. If necessary, briefly centrifuge the vial on a tabletop centrifuge to dislodge any liquid in the container`s cap. Certain products may require to ship with dry ice and additional dry ice fee may apply.
Searchable Terms for HMBS purchase	MBS2562513 is an antibody targeting HMBS. Also known as HMBS Polyclonal Antibody. Reactive in Human, Mouse, Rat. Host: Rabbit.



Immunohistochemistry (IHC) of anti-HMBS antibody
Immunohistochemistry of paraffin-embedded Human thyroid cancer tissue using HMBS Polyclonal Antibody at dilution of 1:25(×200)

ADDITIONAL DETAILS

NCBI/Uniprot data below describe general gene information for HMBS. It may not necessarily be applicable to this product.

NCBI GI #	1170217
NCBI GeneID	3145
NCBI Accession #	P08397.2
NCBI Related Accession #	BC000520
UniProt Primary Accession #	P08397
UniProt Related Accession #	P08397
NCBI Official Full Name	Porphobilinogen deaminase
NCBI Official Synonym Full Names	hydroxymethylbilane synthase
NCBI Official Symbol	HMBS
NCBI Official Synonym Symbols	UPS; PBGD; PORC; PBG-D
UniProt Protein Name	Porphobilinogen deaminase
UniProt Synonym Protein Names	Hydroxymethylbilane synthase; HMBS; Pre-uroporphyrinogen synthase
Protein Family	Porphobilinogen deaminase
UniProt Gene Name	HMBS
UniProt Synonym Gene Names	PBGD; UPS; PBG-D; HMBS
UniProt Entry Name	HEM3_HUMAN
NCBI Protein Information	porphobilinogen deaminase
NCBI Summary for HMBS	This gene encodes a member of the hydroxymethylbilane synthase superfamily. The encoded 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. [provided by RefSeq, Jul 2008]

UniProt Comments for HMBS	HMBS: Tetrapolymerization of the monopyrrole PBG into the hydroxymethylbilane pre-uroporphyrinogen in several discrete steps. Defects in HMBS are the cause of acute intermittent porphyria (AIP). AIP is a form of porphyria. Porphyrias are inherited defects in the biosynthesis of heme, resulting in the accumulation and increased excretion of porphyrins or porphyrin precursors. They are classified as erythropoietic or hepatic, depending on whether the enzyme deficiency occurs in red blood cells or in the liver. AIP is an autosomal dominant form of hepatic porphyria characterized by acute attacks of neurological dysfunctions with abdominal pain, hypertension, tachycardia, and peripheral neuropathy. Most attacks are precipitated by drugs, alcohol, caloric deprivation, infections, or endocrine factors. Belongs to the HMBS family. 2 isoforms of the human protein are produced by alternative splicing. Protein type: EC 2.5.1.61; Cofactor and Vitamin Metabolism - porphyrin and chlorophyll; Transferase; Mitochondrial Chromosomal Location of Human Ortholog: 11q23.3 Cellular Component: cytosol Molecular Function: hydroxymethylbilane synthase activity Biological Process: porphyrin metabolic process; protoporphyrinogen IX biosynthetic process; peptidyl-pyrromethane cofactor linkage; heme biosynthetic process Disease: Porphyria, Acute Intermittent
Research Articles on HMBS	**1.** In the present study, we identified four HMBS gene mutations affecting the ubiquitous isoform of porphobilinogen deaminase (PBGD) and established a dPCR protocol which would be able to detect the different transcripts of this gene.
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