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HMBS Rabbit pAb

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Catalog # AP59129

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	P08397
Predicted	Rat, Pig, Cat, Human, Mouse, Rabbit, Chicken, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	39330
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human HMBS
Epitope Specificity	21-120/361
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SUBCELLULAR LOCATION	Cytoplasm.
DISEASE	Defects in HMBS are the cause of acute intermittent porphyria (AIP) [MIM:176000]. 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.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	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]

Additional Information

Gene ID	3145
Other Names	Porphobilinogen deaminase, PBG-D, 2.5.1.61, Hydroxymethylbilane synthase, HMBS, Pre-uroporphyrinogen synthase, HMBS, PBGD, UPS
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500,ELISA=1:5000-10000

Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

Protein Information

Name	HMBS
Synonyms	PBGD, UPS
Function	As part of the heme biosynthetic pathway, catalyzes the sequential polymerization of four molecules of porphobilinogen to form hydroxymethylbilane, also known as preuroporphyrinogen (PubMed:18004775, PubMed:18936296, PubMed:19138865, PubMed:23815679). Catalysis begins with the assembly of the dipyrromethane cofactor by the apoenzyme from two molecules of porphobilinogen or from preuroporphyrinogen. The covalently linked cofactor acts as a primer, around which the tetrapyrrole product is assembled (PubMed:18936296). In the last step of catalysis, the product, preuroporphyrinogen, is released, leaving the cofactor bound to the holodeaminase intact (PubMed:18936296).
Cellular Location	Cytoplasm, cytosol {ECO:0000250|UniProtKB:P22907}
Tissue Location	[Isoform 1]: Is ubiquitously expressed.

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Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.