



HMBS Polyclonal Antibody

Purified Rabbit Polyclonal Antibody (Pab) **Catalog # AP59129**

Specification

HMBS Polyclonal Antibody - Product Information

Application **Primary Accession** Reactivity Host Clonality

Calculated MW **Physical State** Immunogen

Epitope Specificity

Isotype **Purity**

affinity purified by Protein A

Buffer

SUBCELLULAR LOCATION

SIMILARITY DISEASE

0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.

KLH conjugated synthetic peptide derived

Cytoplasm.

IHC-P, IHC-F, IF, E

Rat, Pig, Cat, Bovine

from human HMBS

P08397

Rabbit

39 KDa

Liquid

laG

Polyclonal

21-120/361

Belongs to the HMBS family. 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.

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

Important Note

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]

HMBS Polyclonal Antibody - Additional Information

Gene ID 3145

Other Names

Porphobilinogen deaminase, PBG-D, 2.5.1.61, Hydroxymethylbilane synthase, HMBS, Pre-uroporphyrinogen synthase, HMBS, PBGD, UPS

Target/Specificity

Isoform 1 is ubiquitously expressed. Isoform 2 is found only in erythroid cells.

Dilution

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<span class ="dilution_IHC-P">IHC-P~~N/A</span><br \> <span class
="dilution_IHC-F">IHC-F~~N/A</span><br \> <span class
="dilution_IF">IF~~1:50~200</span><br \> <span class ="dilution_E">E~~N/A</span>
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Storage

Store at -20 $^{\circ}$ 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 $^{\circ}$ C.

HMBS Polyclonal Antibody - 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.

HMBS Polyclonal Antibody - Protocols







Provided below are standard protocols that you may find useful for product applications.

- Western Blot
- Blocking Peptides
- Dot Blot
- <u>Immunohistochemistry</u>
- <u>Immunofluorescence</u>
- Immunoprecipitation
- Flow Cytomety
- Cell Culture

HMBS Polyclonal Antibody - Images