HMBS Peptide - C-Terminal Region Blocking Peptide

PRODUCT OVERVIEW

Product Name	HMBS, Blocking Peptide
Full Product Name	HMBS Peptide - C-terminal region
Product Gene Name	HMBS blocking peptide
Product Synonym Gene Name	UPS; PBGD; PORC; PBG-D; HMBS
Sequence	Synthetic peptide located within the following region: VPAQHEDGPE DDPQLVGITA RNIPRGPQLA AQNLGISLAN LLLSKGAKNI
Species Reactivity	Human
Form/Format	Lyophilized powder
Preparation and Storage	Add 100ul of sterile PBS. Final peptide concentration is 1 mg/ml in PBS. For longer periods of storage, store at -20 degree C. Avoid repeat freeze-thaw cycles.
Antibody/Peptide Pairs	HMBS peptide (MBS3244605) is used for blocking the activity of HMBS antibody (MBS3219740)
Molecular Weight	39kDa
OMIM	phenotype 609806
Product Categories/Family for HMBS blocking peptide	Peptide
Applications Tested/Suitable for HMBS blocking peptide	Western Blot (WB)
Related Product Information for HMBS blocking peptide	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 HMBS blocking peptide 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	MBS3244605 is a blocking peptide targeting HMBS. Also known as HMBS Peptide - C-terminal region. Reactive in Human.

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
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
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 a Chinese female patient with very typical Acute intermittent porphyria symptoms, a heterozygous mutation of the HMBS gene was identified in the proband and 7 other family members. Genetic sequencing showed a deletion of 55 basepairs (C.1078_1132delGCCCATTAACTGGTTTGTGGGGCACAGATGCCTGGGTTGCTGCTGTCCAGTGCCT) including the stop codon position, leading to frameshift mutation.
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