PAH Recombinant Protein

PRODUCT OVERVIEW

Product Name	PAH, Recombinant Protein
Full Product Name	PAH Protein, Human, Recombinant (415 Asn/Asp, His Tag)
Product Gene Name	PAH recombinant protein
Product Synonym Names	Human PAH/PH Protein (415 Asn/Asp, His Tag); PH Protein, Human; PKU Protein, Human; PKU1 Protein, Human; phenylalanine hydroxylase
Sequence	Met1-Lys45 2, 415Asn/Asp
Host	Baculovirus-Insect Cells
Form/Format	Lyophilized from sterile 20mM Tris, 500mM NaCl, pH8.0, 10% glycerol. Normally 5%-8% trehalose, mannitol and 0.01% Tween80 are added as protectants before lyophilization. Please refer to the specific buffer information in the hard copy of CoA.
Purity/Purification	>70% as determined by SDS-PAGE
Preparation and Storage	Samples are stable for up to twelve months from date of receipt at -20 degree C to -80 degree C. Store it under sterile conditions at -20 degree C to -80 degree C. It is recommended that the protein be aliquoted for optimal storage. Avoid repeated freeze-thaw cycles. In general, recombinant proteins are provided as lyophilized powder which are shipped at ambient temperature. Bulk packages of recombinant proteins are provided as frozen liquid. They are shipped out with blue ice.
Molecular Weight	452
OMIM	phenotype 612349
3D Structure	ModBase 3D Structure for P00439
Species	Human
Endotoxin	<1.0EU per ug of the protein as determined by the LAL method
Predicted N Terminal	Met
Tag	N-His
Protein Construction	A DNA sequence encoding the human PAH (P00439)(Met 1-Lys 452)(415 Asn/Asp) was expressed, with a polyhistidine tag at the N-terminus.
Reconstitution	A hardcopy of COA with reconstitution instruction is sent along with the products. Please refer to it for detailed information.
Related Product Information for PAH recombinant protein	Background: PAH (phenylalanine hydroxylase), also known as PH, belongs to the biopterin-dependent aromatic amino acid hydroxylase family. It contains 1 ACT domain, N-terminal region of PAH is thought to contain allosteric binding sites for phenylalanine and to constitute an "inhibitory" domain that regulates the activity of a catalytic domain in the C-terminal portion of the molecule. In humans, PAH is expressed both in the liver and the kidney, and there is some indication that it may be differentially regulated in these tissues. PAH catalyzes the hydroxylation of the aromatic side-chain of phenylalanine to generate tyrosine. It is one of three members of the pterin-dependent amino acid hydroxylases, a class of monooxygenase that uses tetrahydrobiopterin and a non-heme iron for catalysis. Defects in PAH are the cause of phenylketonuria (PKU). PKU is an autosomal recessive inborn error of phenylalanine metabolism, due to severe phenylalanine hydroxylase deficiency. It is characterized by blood concentrations of phenylalanine persistently above 1200 mumol.

Product References and Citations for PAH recombinant protein	Fitzpatrick PF, et al. (1999) Tetrahydropterin-dependent amino acid hydroxylases. Annu Rev Biochem. 68:355-81.Olsson E, et al. (2011) Formation of the iron-oxo hydroxylating species in the catalytic cycle of aromatic amino acid hydroxylases. Chemistry. 17(13):3746-58.Bassan A, et al. (2003) Mechanism of aromatic hydroxylation by an activated FeIVO core in tetrahydrobiopterin-dependent hydroxylases. Chemistry. 9(17):4055-67.Panay AJ, et al. (2011) Evidence for a high-spin Fe(IV) species in the catalytic cycle of a bacterial phenylalanine hydroxylase. Biochemistry. 50(11):1928-33.Bassan A, et al. (2003) Mechanism of dioxygen cleavage in tetrahydrobiopterin-dependent amino acid hydroxylases. Chemistry. 9(1):106-15.
Other Notes	Small volumes of PAH recombinant protein 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 PAH purchase	MBS8121076 is a recombinant protein targeting PAH. Also known as PAH Protein, Human, Recombinant (415 Asn/Asp, His Tag). Host: Baculovirus-Insect Cells.



SDS-Page of PAH recombinant protein

ADDITIONAL DETAILS

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

NCBI GI #	129973
NCBI GeneID	5053
NCBI Accession #	P00439.1
UniProt Primary Accession #	P00439
UniProt Secondary Accession #	Q16717; Q8TC14
UniProt Related Accession #	P00439
NCBI Official Full Name	Phenylalanine-4-hydroxylase
NCBI Official Synonym Full Names	phenylalanine hydroxylase
NCBI Official Symbol	PAH
NCBI Official Synonym Symbols	PH; PKU; PKU1
UniProt Protein Name	Phenylalanine-4-hydroxylase
UniProt Synonym Protein Names	Phe-4-monooxygenase
Protein Family	Phenylalanine-4-hydroxylase
UniProt Gene Name	PAH
UniProt Synonym Gene Names	PAH
UniProt Entry Name	PH4H_HUMAN
NCBI Protein Information	phenylalanine-4-hydroxylase; phe-4-monooxygenase; phenylalanine 4-monooxygenase
NCBI Summary for PAH	PAH encodes the enzyme phenylalanine hydroxylase that is the rate-limiting step in phenylalanine catabolism. Deficiency of this enzyme activity results in the autosomal recessive disorder phenylketonuria. [provided by RefSeq, Jul 2008]

UniProt Comments for PAH	PAH: phenylalanine hydroxylase that is the rate-limiting step in phenylalanine catabolism. Belongs to the biopterin-dependent aromatic amino acid hydroxylase family. Deficiency of this enzyme activity results in the autosomal recessive disorder phenylketonuria. Protein type: EC 1.14.16.1; Amino Acid Metabolism - phenylalanine, tyrosine and tryptophan biosynthesis; Oxidoreductase Chromosomal Location of Human Ortholog: 12q22-q24.2 Cellular Component: cytosol Molecular Function: amino acid binding; iron ion binding; phenylalanine 4-monooxygenase activity Biological Process: L-phenylalanine catabolic process; catecholamine biosynthetic process; amino acid biosynthetic process; neurotransmitter biosynthetic process Disease: Phenylketonuria
Research Articles on PAH	**1.** Two polymorphic variants of PAH appear to be risk factors for NSCL/P, rs7485331 and rs12425434 in a Polish population.
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