

PAH Protein, Human, Recombinant (415 Asn/Asp, His)

General Information

Synonyms:	phenylalanine hydroxylase;PH;PKU1;PKU
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. Predicted N terminal: Met
Species:	Human
Expression Host:	Baculovirus Insect Cells
Accession:	P00439
Molecular Weight:	54 kDa (predicted); 50 kDa (reducing conditions)

QC Testing

Biological Activity:	Activity testing is in progress. It is theoretically active, but we cannot guarantee it. If you require protein activity, we recommend choosing the eukaryotic expression version first.
Purity:	> 70 % as determined by SDS-PAGE
Endotoxin:	< 1.0 EU/μg of the protein as determined by the LAL method.
Formulation:	Lyophilized from a solution filtered through a 0.22 μm filter, containing 20 mM Tris, 500 mM NaCl, pH 8.0, 10% glycerol. Typically, a mixture containing 5% to 8% trehalose, mannitol, and 0.01% Tween 80 is incorporated as a protective agent before lyophilization.

Preparation and Storage

Reconstitution:
Reconstituted with sterile deionized water to 0.25 mg/mL. Reconstitution conditions may vary depending on the lot.

Stability & Storage:

It is recommended to store recombinant proteins at -20°C to -80°C for future use. Lyophilized powders can be stably stored for over 12 months, while liquid products can be stored for 6-12 months at -80°C. For reconstituted protein solutions, the solution can be stored at -20°C to -80°C for at least 3 months. Please avoid multiple freeze-thaw cycles and store products in aliquots.

Actual storage temperature shall be subject to the COA.

Shipping:

In general, lyophilized powders are shipped with blue ice, while solutions are shipped with dry ice.

Protein Background

PAH (phenylalanine hydroxylase), also known as PH, belongs to the bipterin-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 μmol .

Reference

- 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.
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