

Human PAH / PH Protein (415 Asn/Asp, His Tag)

Catalog Number: 12081-H07B



Sino Biological Inc.
Biological Solution Specialist

General Information

Gene Name Synonym:

PH; PKU; PKU1; AW106920

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.

Source: Human

Expression Host: Baculovirus-Insect Cells

QC Testing

Purity: > 90 % as determined by SDS-PAGE

Endotoxin:

< 1.0 EU per µg of the protein as determined by the LAL method

Stability:

Samples are stable for up to twelve months from date of receipt at -70 °C

Predicted N terminal: Met

Molecular Mass:

The recombinant human PAH consists of 471 amino acids and predicts a molecular mass of 54 kDa. It migrates as an approximately 50 kDa band in SDS-PAGE under reducing conditions.

Formulation:

Lyophilized from sterile 20mM Tris, 500mM NaCl, pH 8.0, 10% glycerol

Normally 5 % - 8 % trehalose, mannitol and 0.01% Tween80 are added as protectants before lyophilization. Specific concentrations are included in the hardcopy of COA. Please contact us for any concerns or special requirements.

Usage Guide

Storage:

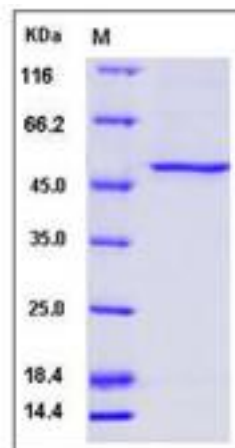
Store it under sterile conditions at -20°C to -80°C upon receiving. Recommend to aliquot the protein into smaller quantities for optimal storage.

Avoid repeated freeze-thaw cycles.

Reconstitution:

Detailed reconstitution instructions are sent along with the products.

SDS-PAGE:



Protein Description

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.

References

1. Fitzpatrick PF, *et al.* (1999) Tetrahydropterin-dependent amino acid hydroxylases. *Annu Rev Biochem.* 68:355-81.
2. 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.
3. Bassan A, *et al.* (2003) Mechanism of aromatic hydroxylation by an activated FeIVO core in tetrahydrobiopterin-dependent hydroxylases. *Chemistry.* 9(17):4055-67.

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