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

Catalog Number:PKSH030912

Note: Centrifuge before opening to ensure complete recovery of vial contents.

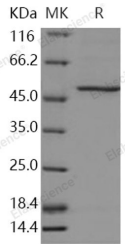
Description

Synonyms	PH;PKU;PKU1
Species	Human
Expression Host	Baculovirus-Insect Cells
Sequence	Met 1-Lys 452
Accession	P00439
Calculated Molecular Weight	54.0 kDa
Observed molecular weight	50 kDa
Tag	N-His

Properties

Purity	> 70 % as determined by reducing SDS-PAGE.
Endotoxin	< 1.0 EU per μg of the protein as determined by the LAL method.
Storage	Generally, lyophilized proteins are stable for up to 12 months when stored at -20 to -80℃. Reconstituted protein solution can be stored at 4-8℃ for 2-7 days. Aliquots of reconstituted samples are stable at < -20℃ for 3 months.
Shipping	This product is provided as lyophilized powder which is shipped with ice packs.
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. Please refer to the specific buffer information in the printed manual.
Reconstitution	Please refer to the printed manual for detailed information.

Data



> 70 % as determined by reducing SDS-PAGE.

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

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phenylalanine hydroxylase deficiency. It is characterized by blood concentrations of phenylalanine persistently above 1200 mumol.

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