



# PAH Monoclonal Antibody

<b>Catalog No</b>	YP-mAb-02732
<b>Isotype</b>	IgG
<b>Reactivity</b>	Human;Mouse;Rat
<b>Applications</b>	WB
<b>Gene Name</b>	PAH
<b>Protein Name</b>	Phenylalanine-4-hydroxylase
<b>Immunogen</b>	The antiserum was produced against synthesized peptide derived from human PAH. AA range:351-400
<b>Specificity</b>	PAH Monoclonal Antibody detects endogenous levels of PAH protein.
<b>Formulation</b>	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
<b>Source</b>	Monoclonal, Mouse,IgG
<b>Purification</b>	The antibody was affinity-purified from mouse antiserum by affinity-chromatography using epitope-specific immunogen.
<b>Dilution</b>	WB 1:500-1:2000
<b>Concentration</b>	1 mg/ml
<b>Purity</b>	≥90%
<b>Storage Stability</b>	-20°C/1 year
<b>Synonyms</b>	PAH; Phenylalanine-4-hydroxylase; PAH; Phe-4-monooxygenase
<b>Observed Band</b>	51kD
<b>Cell Pathway</b>	cytosol,extracellular exosome,
<b>Tissue Specificity</b>	Liver,
<b>Function</b>	catalytic activity:L-phenylalanine + tetrahydrobiopterin + O(2) = L-tyrosine + 4a-hydroxytetrahydrobiopterin.,cofactor:Fe(2+) ion.,disease:Defects in PAH are the cause of hyperphenylalaninemia (HPA) [MIM:261600]. HPA is the mildest form of phenylalanine hydroxylase deficiency.,disease:Defects in PAH are the cause of non-phenylketonuria hyperphenylalaninemia (Non-PKU HPA) [MIM:261600]. Non-PKU HPA is a mild form of phenylalanine hydroxylase deficiency characterized by phenylalanine levels persistently below 600 μmol, which allows normal intellectual and behavioral development without treatment. Non-PKU HPA is usually caused by the combined effect of a mild hyperphenylalaninemia mutation and a severe one.,disease:Defects in PAH are the cause of phenylketonuria (PKU) [MIM:261600]. PKU is an autosomal recessive inborn error of phenylalanine metabolism, due to severe phenylalanine hydroxylase



## Background

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],

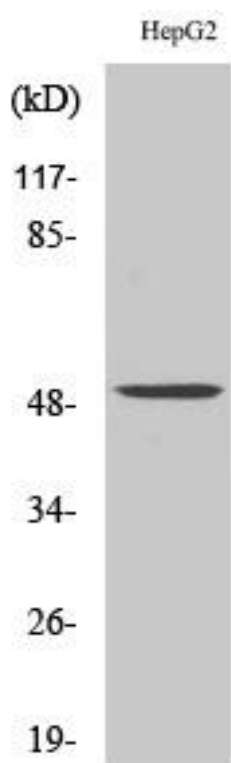
## matters needing attention

Avoid repeated freezing and thawing!

## Usage suggestions

This product can be used in immunological reaction related experiments. For more information, please consult technical personnel.

## Products Images



Western Blot analysis of various cells using PAH Monoclonal Antibody