Best Tools for immunology Research

PAH Polyclonal Antibody

Catalog No :	YT3568
Reactivity :	Human;Mouse;Rat
Applications :	WB;IHC;IF;ELISA
Target :	PAH
Fields :	>>Phenylalanine metabolism;>>Phenylalanine, tyrosine and tryptophan biosynthesis;>>Folate biosynthesis;>>Metabolic pathways;>>Biosynthesis of amino acids
Gene Name :	PAH
Protein Name :	Phenylalanine-4-hydroxylase
Human Gene Id :	5053
Human Swiss Prot No :	P00439
Mouse Gene Id :	18478
Mouse Swiss Prot No :	P16331
Rat Swiss Prot No :	P04176
Immunogen :	The antiserum was produced against synthesized peptide derived from human PAH. AA range:351-400
Specificity :	PAH Polyclonal Antibody detects endogenous levels of PAH protein.
Formulation :	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
Source :	Polyclonal, Rabbit,IgG
Dilution :	WB 1:500 - 1:2000. IHC 1:100 - 1:300. ELISA: 1:40000.. IF 1:50-200
Purification :	The antibody was affinity-purified from rabbit antiserum by affinity-

chromatography using epitope-specific immunogen.

Concentration : 1 mg/ml

Storage Stability : -15°C to -25°C/1 year(Do not lower than -25°C)

Observed Band : 51kD

Cell Pathway : Phenylalanine metabolism;Phenylalanine; tyrosine and tryptophan biosynthesis;

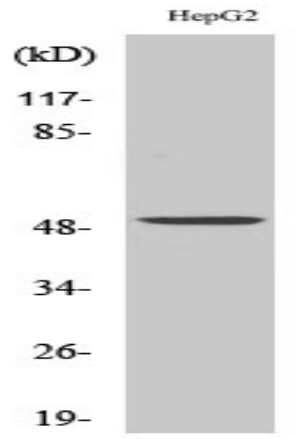
Background : PAH encodes the enzyme phenylalanine hydroxylase that is the rate-limiting step in phenylalanine catabolism. Deficiency of this enzyme activity results in the autosomal recessive disorder phenylketonuria. [provided by RefSeq, Jul 2008],

Function : 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 mumol, 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 hydroxylas

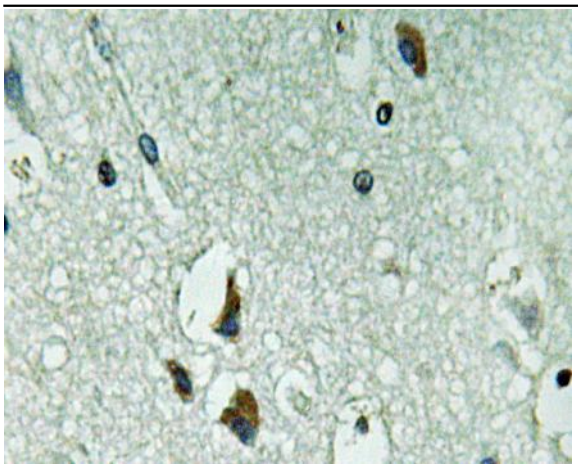
Subcellular Location : cytosol,extracellular exosome,

Expression : Liver,

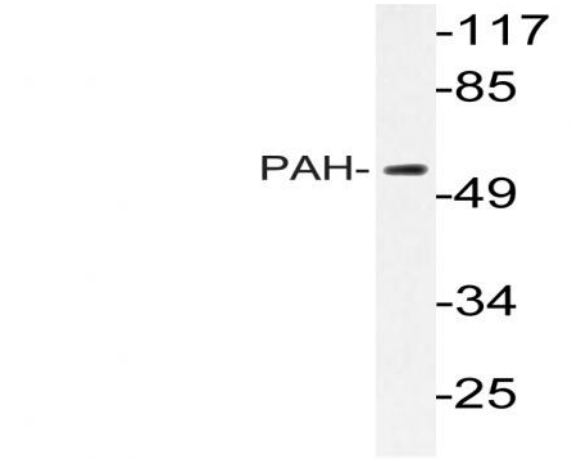
Products Images



Western Blot analysis of various cells using PAH Polyclonal Antibody



Immunohistochemistry analysis of PAH antibody in paraffin-embedded human brain tissue.



Western blot analysis of lysate from HepG2 cells, using PAH antibody.