PAH Rabbit pAb

CatalogNo: YT3568

Key Features

Host Species
- Rabbit

Reactivity
- Human,Mouse,Rat

Applications
- WB,IHC,IF,ELISA

MW
- 51kD (Observed)

Isotype
- IgG

Recommended Dilution Ratios

WB 1:500-1:2000
IHC 1:100-1:300
ELISA 1:40000
IF 1:50-200

Storage

Storage*	-15°C to -25°C/1 year(Do not lower than -25°C)
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.

Basic Information

Clonality	Polyclonal

Immunogen Information

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.

Target Information

Gene name PAH

Protein Name Phenylalanine-4-hydroxylase

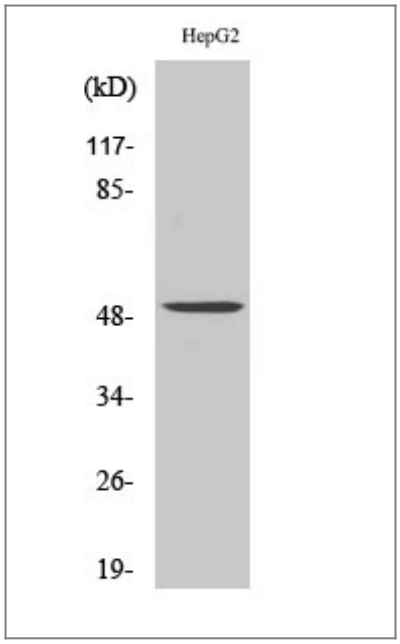
Organism	Gene ID	UniProt ID
Human	5053;	P00439;
Mouse	18478;	P16331;
Rat		P04176;

Cellular Localization cytosol,extracellular exosome,

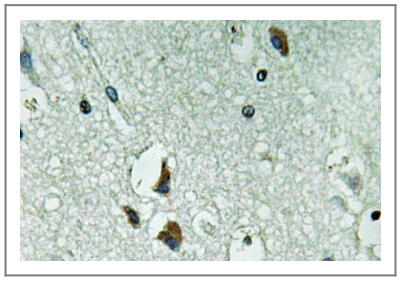
Tissue specificity Liver,

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 hydroxylase deficiency. It is characterized by blood concentrations of phenylalanine persistently above 1200 mumol (normal concentration 100 mumol) which usually causes mental retardation (unless low phenylalanine diet is introduced early in life). They tend to have light pigmentation, rashes similar to eczema, epilepsy, extreme hyperactivity, psychotic states and an unpleasant 'mousy' odor.,enzyme regulation: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.,online information:Phenylalanine hydroxylase entry,online information:Phenylalanine hydroxylase locus knowledgebase,pathway:Amino-acid degradation; L-phenylalanine degradation; acetoacetic acid and fumarate from L-phenylalanine: step 1/6.,polymorphism:The Glu-274 variant occurs on approximately 4% of African-American PAH alleles. The enzyme activity of the variant protein is indistinguishable from that of the wild-type form.,similarity:Belongs to the biopterin-dependent aromatic amino acid hydroxylase family.,similarity:Contains 1 ACT domain.,subunit:Homodimer.,

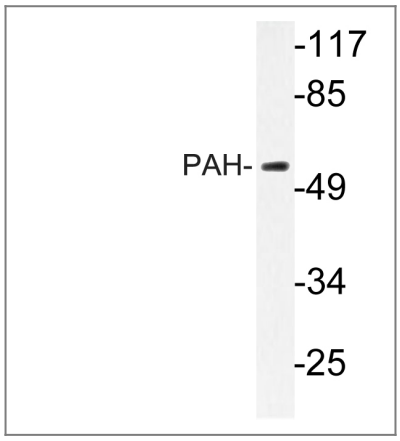
Validation Data



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.

Contact information

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Please scan the QR code to access additional product information:
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