

## PAH Polyclonal Antibody

|                              |                                                                                                                                                           |
|------------------------------|-----------------------------------------------------------------------------------------------------------------------------------------------------------|
| <b>Catalog No :</b>          | YT3568                                                                                                                                                    |
| <b>Reactivity :</b>          | Human;Mouse;Rat                                                                                                                                           |
| <b>Applications :</b>        | WB;IHC;IF;ELISA                                                                                                                                           |
| <b>Target :</b>              | PAH                                                                                                                                                       |
| <b>Fields :</b>              | >>Phenylalanine metabolism;>>Phenylalanine, tyrosine and tryptophan biosynthesis;>>Folate biosynthesis;>>Metabolic pathways;>>Biosynthesis of amino acids |
| <b>Gene Name :</b>           | PAH                                                                                                                                                       |
| <b>Protein Name :</b>        | Phenylalanine-4-hydroxylase                                                                                                                               |
| <b>Human Gene Id :</b>       | 5053                                                                                                                                                      |
| <b>Human Swiss Prot No :</b> | P00439                                                                                                                                                    |
| <b>Mouse Gene Id :</b>       | 18478                                                                                                                                                     |
| <b>Mouse Swiss Prot No :</b> | P16331                                                                                                                                                    |
| <b>Rat Swiss Prot No :</b>   | P04176                                                                                                                                                    |
| <b>Immunogen :</b>           | The antiserum was produced against synthesized peptide derived from human PAH. AA range:351-400                                                           |
| <b>Specificity :</b>         | PAH Polyclonal 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>              | Polyclonal, Rabbit,IgG                                                                                                                                    |
| <b>Dilution :</b>            | WB 1:500 - 1:2000. IHC 1:100 - 1:300. ELISA: 1:40000.. IF 1:50-200                                                                                        |
| <b>Purification :</b>        | 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;

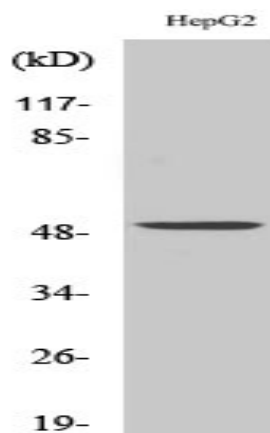
**Background :** PAH encodes the enzyme phenylalanine hydroxylase that is the rate-limiting step in phenylalanine catabolism. Deficiency of this enzyme activity results in the autosomal recessive disorder phenylketonuria. [provided by RefSeq, Jul 2008],

**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 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

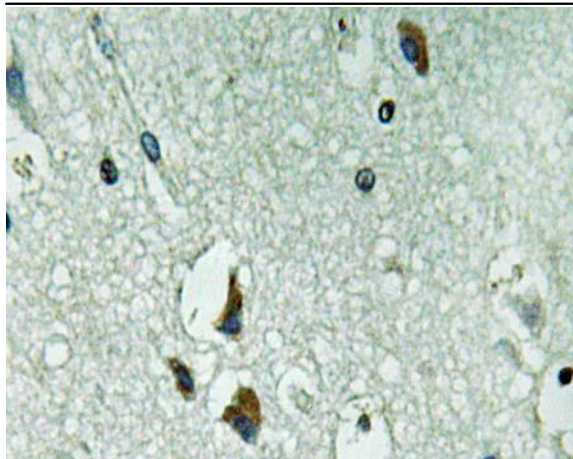
**Subcellular Location :** cytosol, extracellular exosome,

**Expression :** Liver,

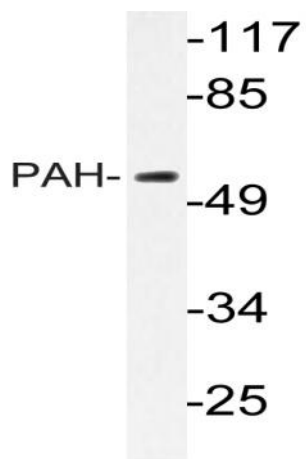
## Products Images



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.