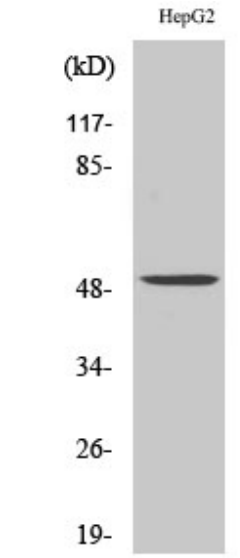
# PAH Polyclonal Antibody

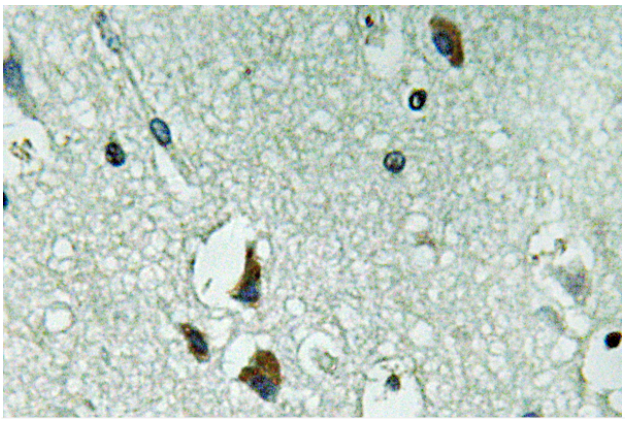
| | |
|---|---|
| **Catalog No** | BYab-02732 |
| **Isotype** | IgG |
| **Reactivity** | Human;Mouse;Rat |
| **Applications** | WB;IHC;IF;ELISA |
| **Gene Name** | PAH |
| **Protein Name** | Phenylalanine-4-hydroxylase |
| **Immunogen** | The antiserum was produced against synthesized peptide derived 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 |
| **Purification** | The antibody was affinity-purified from rabbit antiserum by affinity-chromatography using epitope-specific immunogen. |
| **Dilution** | WB: 1/500 - 1/2000. IHC: 1/100 - 1/300. ELISA: 1/40000.. IF 1:50-200 |
| **Concentration** | 1 mg/ml |
| **Purity** | ≥90% |
| **Storage Stability** | -20°C/1 year |
| **Synonyms** | PAH; Phenylalanine-4-hydroxylase; PAH; Phe-4-monooxygenase |
| **Observed Band** | 51kD |
| **Cell Pathway** | 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 hydroxylas |
| **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. |

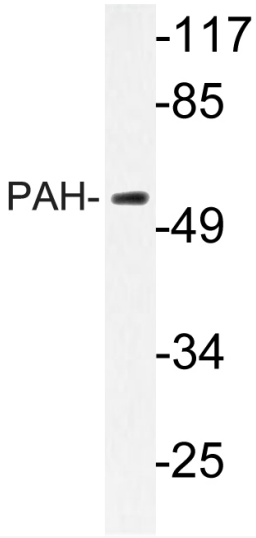
## Products Images




Western Blot analysis of various cells using PAH Polyclonal Antibody



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




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