Product Name: PAH Rabbit Polyclonal Antibody
Catalog #: APRab15696
For research use only.

Summary

Description	Rabbit polyclonal Antibody
Host	Rabbit
Application	WB,IHC,ICC/IF,ELISA
Reactivity	Human,Mouse,Rat
Conjugation	Unconjugated
Modification	Unmodified
Isotype	IgG
Clonality	Polyclonal
Form	Liquid
Concentration	1mg/ml
Storage	Aliquot and store at -20°C (valid for 12 months). Avoid freeze/thaw cycles.
Shipping	Ice bags
Buffer	Liquid in PBS containing 50% glycerol, 0.5% protective protein and 0.02% New type preservative N.
Purification	Affinity purification

Application

Dilution Ratio	WB 1:500-1:2000,IHC 1:100-1:300,ICC/IF 1:50-1:200,ELISA 1:20000-1:40000
Molecular Weight	51kDa

Antigen Information

Gene Name	PAH
Alternative Names	PAH; Phenylalanine-4-hydroxylase; PAH; Phe-4-monooxygenase
Gene ID	5053.0
SwissProt ID	P00439
Immunogen	The antiserum was produced against synthesized peptide derived from human PAH. AA range:351-400

Background

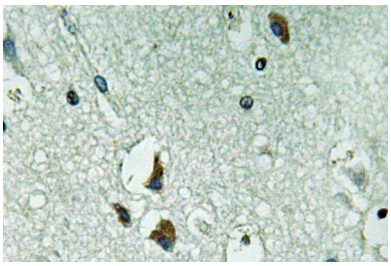
PAH encodes the enzyme phenylalanine hydroxylase that is the rate-limiting step in phenylalanine catabolism. Deficiency of

this enzyme activity results in the autosomal recessive disorder phenylketonuria. [provided by RefSeq, Jul 2008],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.,

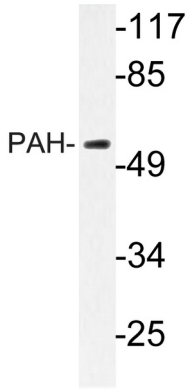
Research Area

Phenylalanine metabolism;Phenylalanine; tyrosine and tryptophan biosynthesis;

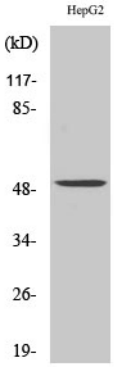
Image Data



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



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



Western Blot analysis of various cells using PAH Polyclonal Antibody