

产品名称: PAH Rabbit Polyclonal Antibody
产品货号: AP Rab15696



产品概述 (Summary)

产品名称 (Production Name)	PAH Rabbit Polyclonal Antibody
描述 (Description)	Rabbit polyclonal Antibody
宿主 (Host)	Rabbit
应用 (Application)	WB,IHC,ICC/IF,ELISA
种属反应性 (Reactivity)	Human,Mouse,Rat

产品性能 (Performance)

偶联物 (Conjugation)	Unconjugated
修饰 (Modification)	Unmodified
同种型 (Isotype)	IgG
克隆 (Clonality)	Polyclonal
形式 (Form)	Liquid
存放说明 (Storage)	Store at 4°C short term. Aliquot and store at -20°C long term. Avoid freeze/thaw cycles.
储存溶液 (Buffer)	Liquid in PBS containing 50% glycerol, 0.5% protective protein and 0.02% New type preservative N.
纯化方式 (Purification)	Affinity purification

免疫原信息 (Immunogen)

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

产品应用 (Application)

稀释比 (Dilution Ratio)	WB 1:500-1:2000,IHC 1:100-1:300,ICC/IF 1:50-1:200,ELISA 1:20000-1:40000
蛋白分子量 (Molecular Weight)	51kDa

研究背景 (Background)

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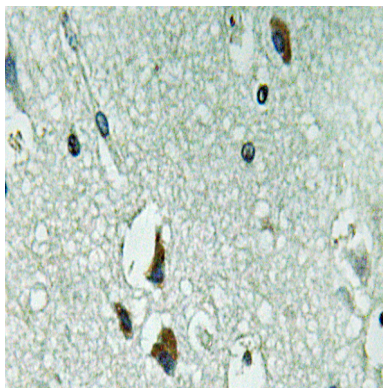


PAH encodes the enzyme phenylalanine hydroxylase that is the rate-limiting step in phenylalanine catabolism. Deficiency of this enzyme activity results in the autosomal recessive disorder phenylketonuria. [provided by RefSeq, Jul 2008], catalytic activity: L-phenylalanine + tetrahydrobiopterin + O(2) = 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 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 deficiency. It is characterized by blood concentrations of phenylalanine persistently above 1200 μmol (normal concentration 100 μmol) 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.,

研究领域 (Research Area)

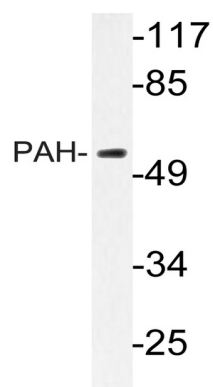
Phenylalanine metabolism; Phenylalanine; tyrosine and tryptophan biosynthesis;

图片 (Image Data)

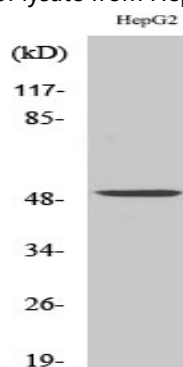


Immunohistochemistry analysis of PAH antibody in paraffin-embedded human brain tissue.

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



Western Blot analysis of various cells using PAH Polyclonal Antibody

注意事项 (Note)

For research use only .