G-Biosciences Image not found or type unknown

Immunotag™ PAH Polyclonal Antibody

Antibody Specification

Catalog No.	ITT3568
Product Description	Immunotag™ PAH Polyclonal Antibody
Size	50 µg, 100 µg
Conjugation	HRP, Biotin, FITC, Alexa Fluor® 350, Alexa Fluor® 405, Alexa Fluor® 488, Alexa Fluor® 555, Alexa Fluor® 594, Alexa Fluor® 647
IMPORTANT NOTE	This product is custom manufactured with a lead time of 3-4 weeks. Once in production, this item cannot be cancelled from an order and is not eligible for return.
Target Protein	PAH
Clonality	Polyclonal
Storage/Stability	-20°C/1 year
Application	WB,IHC-p,ELISA
Recommended Dilution	Western Blot: 1/500 - 1/2000. Immunohistochemistry: 1/100 - 1/300. ELISA: 1/40000. Not yet tested in other applications.
Concentration	1 mg/ml
Reactive Species	Human,Mouse,Rat
Host Species	Rabbit
Immunogen	Synthesized peptide derived from the C-terminal region of human PAH
Specificity	PAH Polyclonal Antibody detects endogenous levels of PAH protein.
Purification	The antibody was affinity-purified from rabbit antiserum by affinity-chromatography using epitope-specific immunogen
Form	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
Gene Name	PAH
Accession No.	P00439 P16331 P04176
Alternate Names	PAH; Phenylalanine-4-hydroxylase; PAH; Phe-4-monooxygenase
Description	phenylalanine hydroxylase(PAH) Homo sapiens 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],

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Cell Pathway/ Category	Phenylalanine metabolism,Phenylalanine, tyrosine and tryptophan biosynthesis,
Protein Expression	Liver,
Subcellular Localization	cytosol,extracellular exosome,
Protein 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.,
Usage	For Research Use Only! Not for diagnostic or therapeutic procedures.

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