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Rabbit Anti-PAH/PH4H antibody

SL21005R

Product Name:	PAH/PH4H
Chinese Name:	苯丙氨酸羟化酶4抗体
Alias:	PAH; pah; PH; PH4H_HUMAN; Phe 4 monooxygenase; Phe-4-monooxygenase; Phenylalanine 4 hydroxylase; Phenylalanine hydroxylase; Phenylalanine-4-hydroxylase; PKU; PKU1.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Cow,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	52kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human PAH/PH4H:1-100/452
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
PubMed:	PubMed
Product Detail:	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] **Subunit:**

Homodimer and homotetramer.

Tissue Specificity:
Optimum temperature is 50 degrees Celsius.

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.
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.
Defects in PAH are the cause of hyperphenylalaninemia (HPA) [MIM:261600]. HPA is the mildest form of phenylalanine hydroxylase deficiency.

Similarity:
Belongs to the biopterin-dependent aromatic amino acid hydroxylase family. Contains 1 ACT domain.

SWISS:
P00439

Gene ID:
5053

Database links:

Entrez Gene: 408024 Chicken

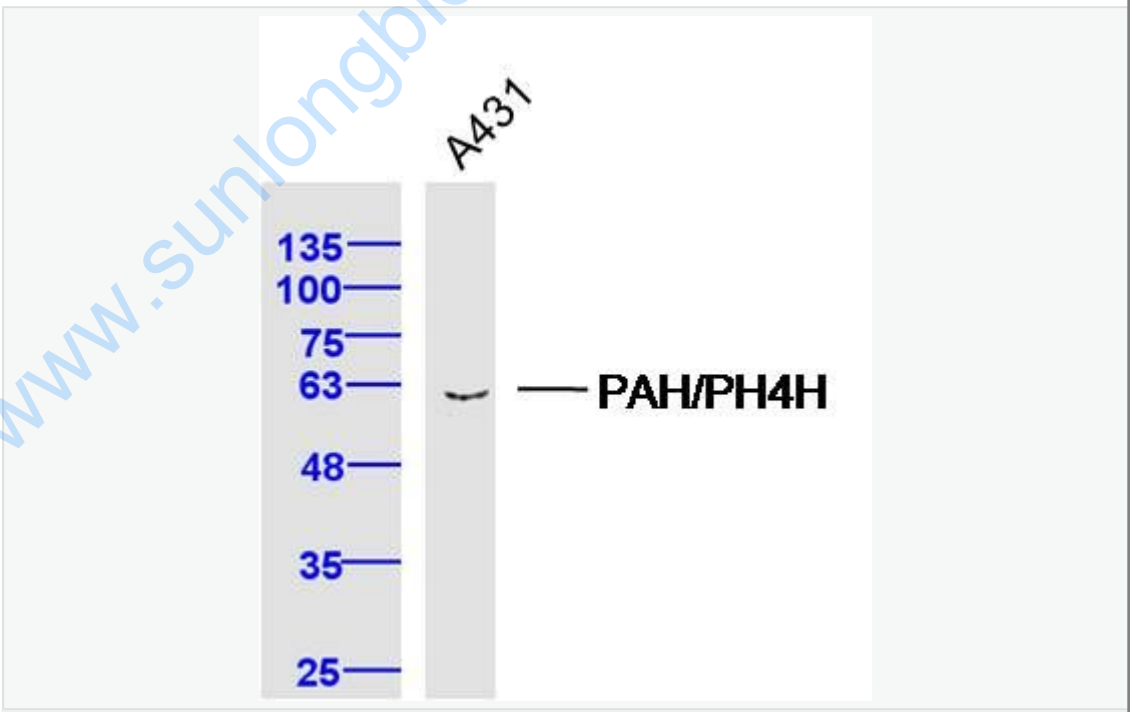
Entrez Gene: 510583 Cow

Entrez Gene: 475446 Dog

Entrez Gene: 5053 Human

Entrez Gene: 18478 Mouse

Entrez Gene: 24616 Rat

	Omim: 612349 Human SwissProt: Q2KIH7 Cow SwissProt: P00439 Human SwissProt: P16331 Mouse SwissProt: P04176 Rat Unigene: 603740 Human Unigene: 263539 Mouse Unigene: 1652 Rat **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Picture:	 Sample: A431 Cell (Human) Lysate at 40 ug Primary: Anti-PAH/PH4H (SL21005R) at 1/300 dilution

	Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution Predicted band size: 52 kD Observed band size: 60 kD