

Mouse Anti-PAH antibody

SLM-51585M

Product Name	PAH
Chinese Name	苯丙氨酸羟化酶 4 单克隆抗体
Alias	PH; PH4H_HUMAN; Phe 4 monooxygenase; Phe-4-monooxygenase; Phenylalanine 4 hydroxylase; Phenylalanine hydroxylase; Phenylalanine-4-hydroxylase; PKU; PKU1.
Research Area	Tumour Cell biology Signal transduction Stem cells Kinases and Phosphatases
Immunogen Species	Mouse
Clonality	Monoclonal
Clone NO.	U8Y5
React Species	Human,Mouse,Rat WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500 (Paraffin sections need antigen repair)
Applications	not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Theoretical molecular weight	52kDa
Cellular localization	cytoplasmic
Form	Liquid
Concentration	1mg/ml
immunogen	KLH conjugated synthetic peptide derived from human PAH: 121-300/452
Lsotype	IgG1, κ
Purification	affinity purified by Protein G
Buffer Solution	1M TBS(pH7.4) with 1% BSA, 3% Proclin300 and 50% Glycerol.
Storage	Shipped at 4°C. Store at -20 °C for one year. Avoid repeated freeze/thaw cycles.
Attention	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
PubMed	PubMed

This gene encodes a member of the bipterin-dependent aromatic amino acid hydroxylase protein family. The encoded phenylalanine hydroxylase enzyme hydroxylates phenylalanine to tyrosine and is the rate-limiting step in phenylalanine catabolism. Deficiency of this enzyme activity results in the autosomal recessive disorder phenylketonuria. [provided by RefSeq, Aug 2017]

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 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.

Product Detail

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.

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

Similarity:

Belongs to the bipterin-dependent aromatic amino acid hydroxylase family. Contains 1 ACT domain.

SWISS:

P00439

Gene ID:

5053

Database links:

[Entrez Gene: 5053](#) Human

[Entrez Gene: 18478](#) Mouse

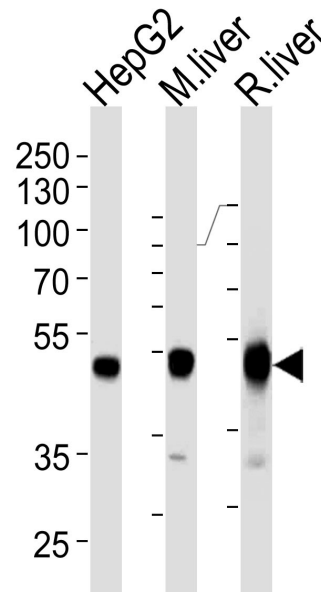
[Entrez Gene: 24616](#) Rat

[SwissProt: P00439](#) Human

[SwissProt: P16331](#) Mouse

[SwissProt: P04176](#) Rat

Product Picture



Sample:

Lane 1: HepG2 cell lysates

Lane 2: Mouse liver tissue lysates

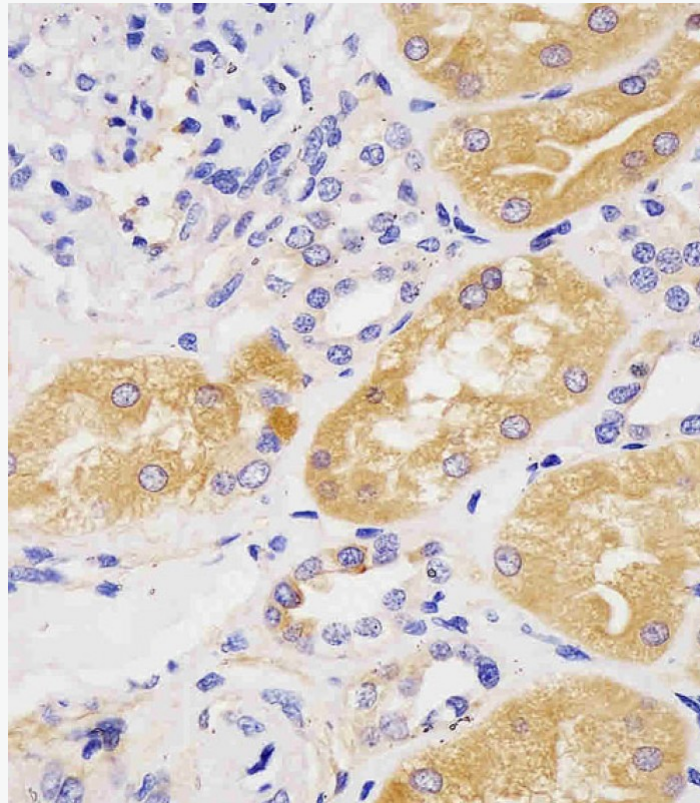
Lane 3: Rat liver tissue lysates

Primary: Anti-PAH (SLM-51585M) at 1/1000 dilution

Secondary: IRDye800CW Goat Anti-Mouse IgG at 1/20000 dilution

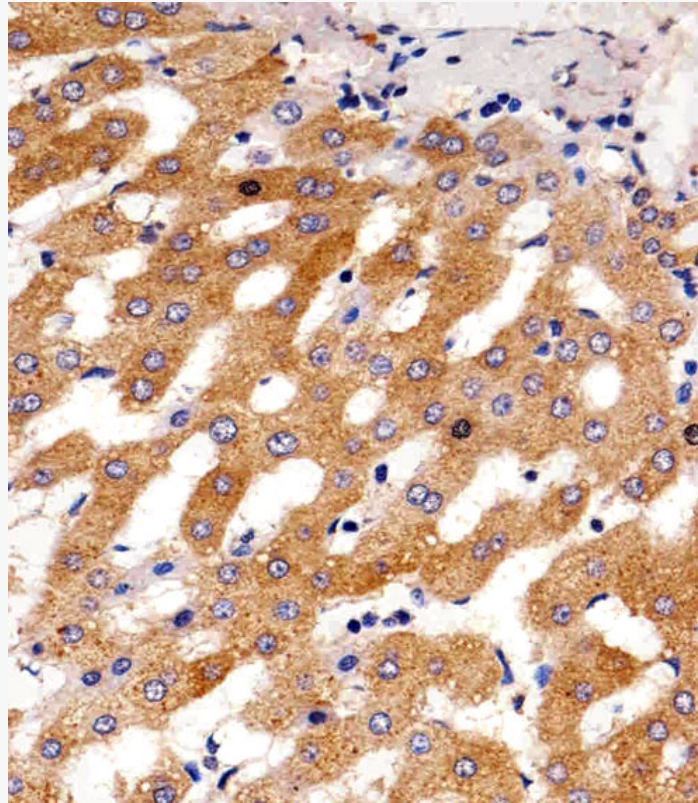
Predicted band size: 32 kD

Observed band size: 30 kD



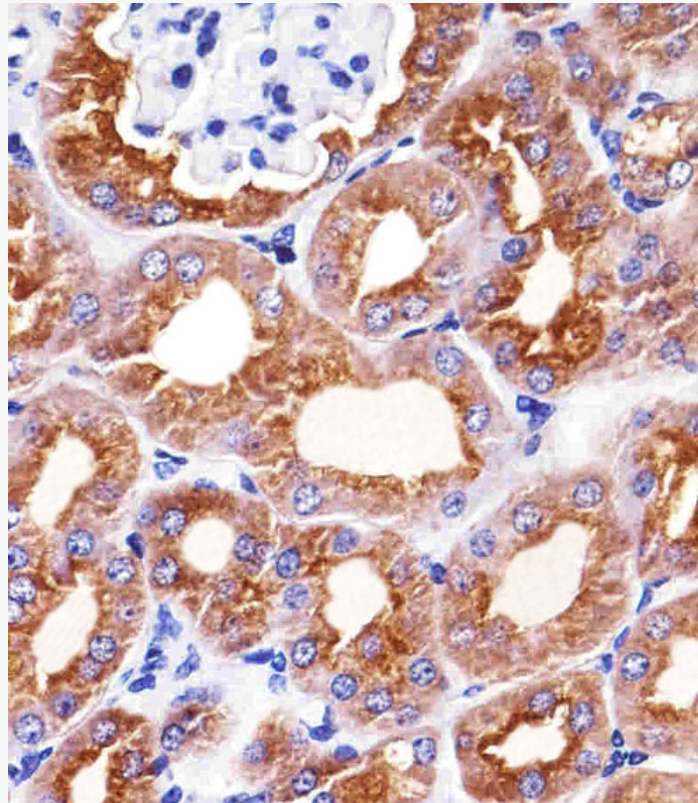
Paraformaldehyde-fixed, paraffin embedded (human kidney tissue sections); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min;

Antibody incubation with (PAH) Monoclonal Antibody, Unconjugated (SLM-51585M) at 1:200 overnight at 4°C, followed by operating according to SP Kit(Mouse)(sp-0024) instructions and DAB staining.



Paraformaldehyde-fixed, paraffin embedded (Human liver section);
Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min;
Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes;
Blocking buffer (normal goat serum) at 37°C for 30min; Antibody
incubation with (PAH) Monoclonal Antibody, Unconjugated
(SLM-51585M) at 1:200 overnight at 4°C, followed by operating

according to SP Kit(Mouse)(sp-0024) instructions and DAB staining.



Paraformaldehyde-fixed, paraffin embedded (Human kidney); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (PAH) Monoclonal Antibody, Unconjugated (SLM-51585M) at 1:200 overnight at 4°C, followed by operating according to SP Kit(Mouse)(sp-0024) instructions and DAB staining.