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Rabbit Anti-Fumarylacetoacetate hydrolase antibody

SL16194R

Product Name:	Fumarylacetoacetate hydrolase
Chinese Name:	延胡索酰乙酰乙酸水解酶抗体
Alias:	Beta diketonase; FAA; FAAA_HUMAN; FAH; Fumarylacetoacetase; Fumarylacetoacetate; Fumarylacetoacetate hydrolase.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Pig,
Applications:	ELISA=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:	47kDa
Cellular localization:	cytoplasmicExtracellular matrixSecretory protein
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human Fumarylacetoacetate hydrolase:21-120/419
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:	This gene encodes the last enzyme in the tyrosine catabolism pathway. FAH deficiency is associated with Type 1 hereditary tyrosinemia (HT). [provided by RefSeq, Jul 2008]

Function:
Fumarylacetoacetate hydrolase is the last enzyme in the tyrosine catabolism pathway. FAH deficiency is associated with Type 1 hereditary tyrosinemia (HT). This is an autosomal recessive inborn error of metabolism that occurs in both an acute and a chronic form. Clinical characteristics of the acute form include hepatic failure and death in infancy, whereas children with the chronic form have renal tubular dysfunction and hypophosphatemic rickets, progressive liver disease with development of hepatocellular carcinoma. Dietary treatment with restriction of tyrosine and phenylalanine alleviates the rickets, but liver transplantation has so far been the only definite treatment.

Subunit:
Homodimer.

Tissue Specificity:
Mainly expressed in liver and kidney. Lower levels are also detected in many other tissues.

Similarity:
Belongs to the FAH family.

SWISS:
P16930

Gene ID:
2184

Database links:

Entrez Gene: 2184 Human

Entrez Gene: 14085 Mouse

Entrez Gene: 29383 Rat

Omim: 613871 Human

SwissProt: P16930 Human

SwissProt: P35505 Mouse

SwissProt: P25093 Rat

Important Note:
This product as supplied is intended for research use only, not for use in human,

	therapeutic or diagnostic applications.