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[Primary Antibody]

Rabbit Anti-Fumarylacetoacetate hydrolase antibody

Catalog Number: SL16194R

FOR RESEARCH USE ONLY

[DATASHEET]

Host:	Rabbit
Isotype:	IgG
Clonality:	Polyclonal
Target:	Fumarylacetoacetate hydrolase
Immunogen:	KLH conjugated synthetic peptide derived from human Fumarylacetoacetate hydrolase: 21-120/419
Purification:	affinity purified by Protein A
Concentration:	1mg/ml
Storage:	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol. Shipped at 4℃. Store at -20 °C for one year. Avoid repeated freeze/thaw cycles.
Applications:	IHC-P=1:100-500 IHC-F=1:100-500 ICC/IF=1:100-500 IF=1:100-500 ELISA=1:5000-10000 （Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Reactivity:	(predicted: Human, Mouse, Rat, Pig)
Predicted MW.:	47kDa
Subcellular Location:	cytoplasmic Extracellular matrix Secretory protein
Background:	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

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	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. **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

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