10320 Camino Santa Fe, Suite G
San Diego, CA 92121
Tel: 858.875.1900 Fax: 858.875.1999

FECH Rabbit pAb

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Catalog # AP59280

Product Information

Application	WB, IHC-P, IHC-F, IF
Primary Accession	P22830
Predicted	Rat, Pig, Dog, Human, Mouse, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	47862
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human EPB41
Epitope Specificity	81-180/423
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SUBCELLULAR LOCATION	Mitochondrion inner membrane; Peripheral membrane protein; Matrix side.
DISEASE	Defects in FECH are the cause of erythropoietic protoporphyria (EPP) [MIM:177000]. Porphyrias are inherited defects in the biosynthesis of heme, resulting in the accumulation and increased excretion of porphyrins or porphyrin precursors. They are classified as erythropoietic or hepatic, depending on whether the enzyme deficiency occurs in red blood cells or in the liver. EPP is a form of porphyria marked by excessive protoporphyrin in erythrocytes, plasma, liver and feces, and by widely varying photosensitive skin changes ranging from a burning or pruritic sensation to erythema, edema and wheals.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	Ferrochelatase catalyzes the insertion of the ferrous form of iron into protoporphyrin IX in the heme synthesis pathway, and is localised in the mitochondrion. Defects in ferrochelatase are associated with protoporphyria. Two transcript variants encoding different isoforms have been found for this gene. Porphyrias are a group of inherited or acquired disorders of certain enzymes in the heme biosynthetic pathway (also called porphyrin pathway). They are broadly classified as hepatic porphyrias or erythropoietic porphyrias, based on the site of the overproduction and mainly accumulation of the porphyrins (or their chemical precursors). They manifest with either skin problems, or neurological complications, or occasionally both.

Additional Information

Gene ID	2235
Other Names	Ferrochelatase, mitochondrial, 4.98.1.1, Heme synthase, Protoheme ferro-lyase, FECH (HGNC:3647)

Dilution	WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500,ELISA=1:5000-10000
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. 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.

Protein Information

Name	FECH (HGNC:3647)
Function	Catalyzes the ferrous insertion into protoporphyrin IX and participates in the terminal step in the heme biosynthetic pathway.
Cellular Location	Mitochondrion inner membrane {ECO:0000250|UniProtKB:P22315}; Peripheral membrane protein {ECO:0000250|UniProtKB:P22315}; Matrix side {ECO:0000250|UniProtKB:P22315}

Background

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