SunLong Biotech Co.,LTD
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Rabbit Anti-FH antibody

SL3938R

Product Name	FH
Chinese Name	富马酸水合酶抗体
Alias	mitochondrial; Fumarase; Fumarate hydratase; Fumarate hydratase mitochondrial; FUMH_HUMAN; HLRCC; LRCC; MCL; MCUL 1; MCUL1; Multiple hereditary cutaneous leiomyomata.
Research Area	Tumour Cell biology immunology transcriptional regulatory factor Mitochondrion
Immunogen Species	Rabbit
Clonality	Polyclonal
React Species	Mouse,Rat(predicted:Human,Dog,Pig,Cow,Horse,Rabbit,Sheep)
Applications	WB=1:500-2000 （Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Theoretical molecular weight	51kDa
Cellular localization	cytoplasmic Mitochondrion
Form	Liquid
Concentration	1mg/ml
immunogen	KLH conjugated synthetic peptide derived from human Fumarate hydratase: 411-510/510
Lsotype	IgG
Purification	affinity purified by Protein A
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
Product Detail	The protein encoded by this gene is an enzymatic component of the

tricarboxylic acid (TCA) cycle, or Krebs cycle, and catalyzes the formation of L-malate from fumarate. It exists in both a cytosolic form and an N-terminal extended form, differing only in the translation start site used. The N-terminal extended form is targeted to the mitochondrion, where the removal of the extension generates the same form as in the cytoplasm. It is similar to some thermostable class II fumarases and functions as a homotetramer. Mutations in this gene can cause fumarase deficiency and lead to progressive encephalopathy. [provided by RefSeq, Jul 2008]

Function:
Also acts as a tumor suppressor.

Subunit:
Homotetramer.

Subcellular Location:
Cytoplasm, Mitochondrion

Tissue Specificity:
Expressed in red blood cells; underexpressed in red blood cells (cytoplasm) of patients with hereditary non-spherocytic hemolytic anemia of unknown etiology.

DISEASE:
Fumarase deficiency (FMRD): The disease is caused by mutations affecting the gene represented in this entry. A severe autosomal recessive metabolic disorder characterized by early-onset hypotonia, profound psychomotor retardation, and brain abnormalities, such as agenesis of the corpus callosum, gyral defects, and ventriculomegaly. Many patients show neonatal distress, metabolic acidosis, and/or encephalopathy.
Hereditary leiomyomatosis and renal cell cancer (HLRCC): The disease is caused by mutations affecting the gene represented in this entry. A disorder characterized by predisposition to cutaneous and uterine leiomyomas, and papillary type 2 renal cancer which occurs in about 20% of patients.

Similarity:
Belongs to the class-II fumarase/aspartase family. Fumarase subfamily.

SWISS:
P07954

Gene ID:
2271

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Database links:

Entrez Gene: 520260 Cow

Entrez Gene: 480092 Dog

Entrez Gene: 2271 Human

Entrez Gene: 14194 Mouse

Entrez Gene: 100627128 Pig

Entrez Gene: 24368 Rat

Entrez Gene: 393938 Zebrafish

Omim: 136850 Human

SwissProt: P07954 Human

SwissProt: P97807 Mouse

SwissProt: P10173 Pig

SwissProt: P14408 Rat

SwissProt: Q7SX99 Zebrafish

Unigene: 592490 Human

Unigene: 41502 Mouse

Unigene: 29782 Rat

Unigene: 104452 Zebrafish

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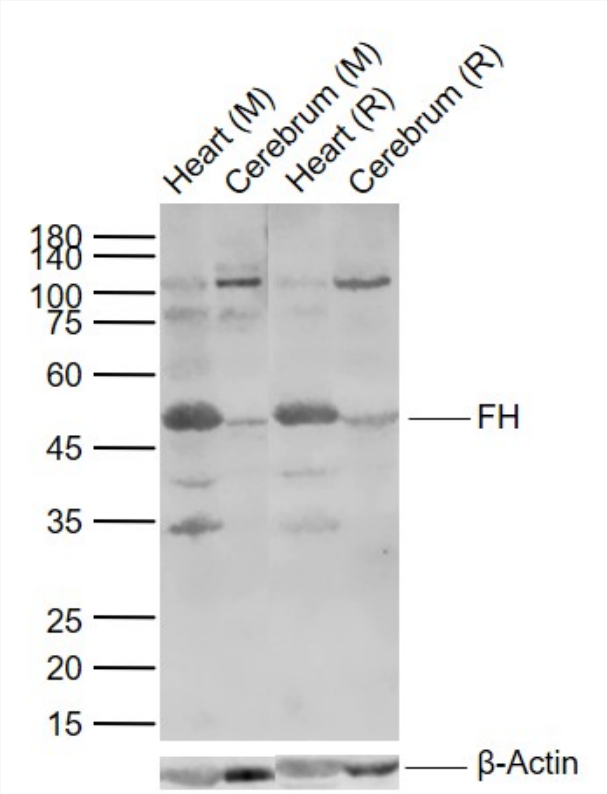
Product Picture



Sample:

Lane 1: Mouse Heart tissue lysates

Lane 2: Mouse Cerebrum tissue lysates

Lane 3: Rat Heart tissue lysates

Lane 4: Rat Cerebrum tissue lysates

Primary: Anti-FH (SL3938R) at 1/1000 dilution

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

Predicted band size: 51 kDa

Observed band size: 51 kDa