

FH Polyclonal Antibody

Catalog No: YN5599

Reactivity: Human; Mouse; Rat

Applications: WB

Target: FH

Fields: >>Citrate cycle (TCA cycle);>>Pyruvate metabolism;>>Metabolic

pathways;>>Carbon metabolism;>>Cushing syndrome;>>Pathways in

cancer;>>Renal cell carcinoma

Gene Name: FH

Protein Name : Fumarate hydratase, mitochondrial

P07954

P97807

Human Gene Id: 2271

Human Swiss Prot

No:

Mouse Gene Id: 14194

Mouse Swiss Prot

No:

Rat Swiss Prot No: P14408

Immunogen: Recombinant Protein of Fumarate hydratase, mitochondrial

Specificity: The antibody detects endogenous fumarase proteins.

Formulation : PBS, pH 7.4, containing 0.5%BSA, 0.02% sodium azide as Preservative and

50% Glycerol.

Source: Polyclonal, Rabbit, lgG

Dilution: WB 1:1000

Purification: The antibody was affinity-purified from rabbit antiserum by affinity-

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Host:

Modifications:

Rabbit

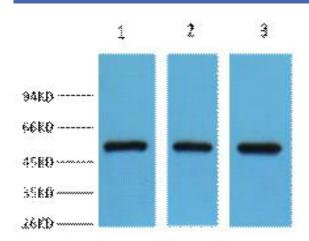
Unmodified

chromatography using epitope-specific immunogen. -15°C to -25°C/1 year(Do not lower than -25°C) **Storage Stability:** Observed Band: 50kD Citrate cycle (TCA cycle);Pathways in cancer;Renal cell carcinoma; **Cell Pathway:** The protein encoded by this gene is an enzymatic component of the tricarboxylic **Background:** 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:** catalytic activity:(S)-malate = fumarate + H(2)O.,disease:Defects in FH are the cause of fumarase deficiency (FD) [MIM:606812]; also known as fumaricaciduria. FD is characterized by progressive encephalopathy, developmental delay, hypotonia, cerebral atrophy and lactic and pyruvic acidemia., disease: Defects in FH are the cause of hereditary leiomyomatosis and renal cell cancer (HLRCC) [MIM:605839]., disease: Defects in FH are the cause of multiple cutaneous and uterine leiomyomata (MCUL1) [MIM:150800]. MCUL1 is an autosomal dominant condition in which affected individuals develop benign smooth muscle tumors (leiomyomata) of the skin. Affected females also usually develop leiomyomata of the uterus (fibroids).,function:Also acts as a tumor suppressor., miscellaneous: There are 2 substrate binding sites: the catalytic A site, and the non-catalytic B site that may play a role in the transfer of s Subcellular [Isoform Mitochondrial]: Mitochondrion .; [Isoform Cytoplasmic]: Cytoplasm, cytosol . Nucleus . Chromosome . Translocates to the nucleus in response to DNA Location: damage: localizes to DNA double-strand breaks (DSBs) following phosphorylation by PRKDC... **Expression:** Expressed in red blood cells; underexpressed in red blood cells (cytoplasm) of patients with hereditary non-spherocytic hemolytic anemia of unknown etiology. Sort: 17439 No4:

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Western blot analysis of 1) Hela, 2) Mouse Brain, 3) HepG2, diluted at 1:2000. Secondary antibody(catalog#:RS0002) was diluted at 1:20000