Best Tools for immunology Research

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
Human Gene Id :	2271
Human Swiss Prot No :	P07954
Mouse Gene Id :	14194
Mouse Swiss Prot No :	P97807
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,IgG
Dilution :	WB 1:1000
Purification :	The antibody was affinity-purified from rabbit antiserum by affinity-

chromatography using epitope-specific immunogen.

Storage Stability : -15°C to -25°C/1 year(Do not lower than -25°C)

Observed Band : 50kD

Cell Pathway : Citrate cycle (TCA cycle);Pathways in cancer;Renal cell carcinoma;

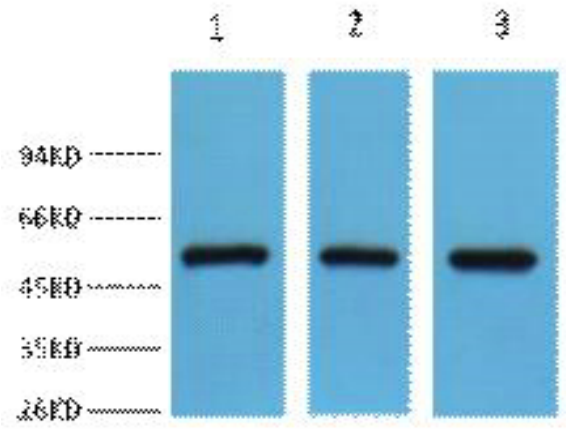
Background : The protein encoded by this gene is an enzymatic component of the tricarboxylic acid (TCA) cycle, or Krebs cycle, and catalyzes the formation of L-malate from fumarate. It exists in both a cytosolic form and an N-terminal extended form, differing only in the translation start site used. 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 : 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 Location : [Isoform Mitochondrial]: Mitochondrion .; [Isoform Cytoplasmic]: Cytoplasm, cytosol . Nucleus . Chromosome . Translocates to the nucleus in response to DNA 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.

Products Images



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