



FH Rabbit mAb

Catalog No	YP-rAb-16891
Isotype	IgG
Reactivity	Human,Mouse,Rat
Applications	WB,IHC,IF,IP,ELISA
Gene Name	FH
Protein Name	Fumarate hydratase, mitochondrial
Purification Process	Protein A
Specificity	Endogenous
Formulation	PBS, 50% glycerol, 0.05% Proclin 300, 0.05%BSA
Source	Monoclonal, Rabbit,IgG
Dilution	IHC 1:200-1:1000; WB 1:2000-1:10000; IF 1:200-1:1000; ELISA 1:5000-1:20000; IP 1:50-1:200; Note: For IHC, we suggest antigen retrieval with TE buffer pH 9.0
Concentration	0.5 mg/ml
Purity	≥90%
Storage Stability	-15 ° C to -25 ° C/1 year(Do not lower than -25 ° C)
Synonyms	FH;Fumarate hydratase, mitochondrial;Fumarate hydratase, mitochondrial ; Fumarase
Observed Band	49kD
Calculated Molecular Weight	55kD
Cell Pathway	[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. .
Tissue Specificity	Expressed in red blood cells; underexpressed in red blood cells (cytoplasm) of patients with hereditary non-spherocytic hemolytic anemia of unknown etiology.
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 substrate or product between the active site and the solvent. Alternatively, the B site may bind allosteric effectors. pathway: Carbohydrate metabolism; tricarboxylic acid cycle. PTM: Isoform Cytoplasmic is acetylated at position 2. similarity: Belongs to the class-II fumarase/aspartase family. Fumarase subfamily. subunit: Homotetramer.

Background

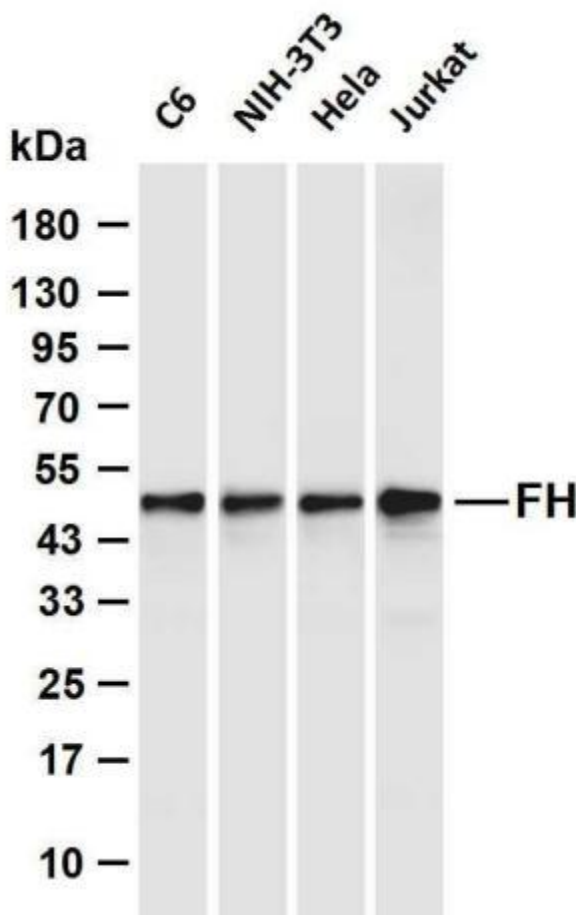
The protein encoded by this gene is an enzymatic component of the tricarboxylic acid (TCA) cycle, or Krebs cycle, and catalyzes the formation of L-malate from fumarate. It exists in both a cytosolic form and an N-terminal extended form, differing only in the translation start site used. The N-terminal 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],

matters needing attention

Avoid repeated freezing and thawing!

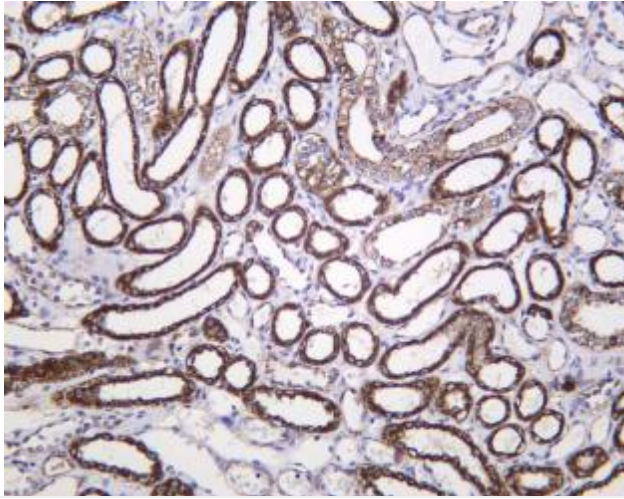
Usage suggestions

This product can be used in immunological reaction related experiments. For more information, please consult technical personnel.



Various whole cell lysates were separated by 4-20% SDS-PAGE, and the membrane was blotted with anti-FH antibody. The HRP-conjugated Goat anti-Rabbit IgG (H + L) antibody was used to detect the antibody. Lane 1: C6 Lane 2: NIH-3T3 Lane 3: HeLa Lane 4: Jurkat Predicted band size: 55kDa Observed band size: 49kDa





Human kidney was stained with anti-FH Rabbit antibody

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