



FH Monoclonal Antibody

Catalog No	YP-mAb-03007
Isotype	IgG
Reactivity	Human;Mouse;Rat
Applications	WB
Gene Name	FH
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	Monoclonal, Mouse,IgG
Purification	The antibody was affinity-purified from mouse antiserum by affinity-chromatography using epitope-specific immunogen.
Dilution	WB 1:500-1:2000
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	EC:4.2.1.2;Fumarase;HsFH;FH;Fumarate hydratase;mitochondrial;mitochondrial (Fumarase) (EC 4.2.1.2); mitochondrial; mitochondrial (Fumarase) (EC 4.2.1.2)
Observed Band	50kD
Calculated Molecular Weight	50kD
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 s

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],

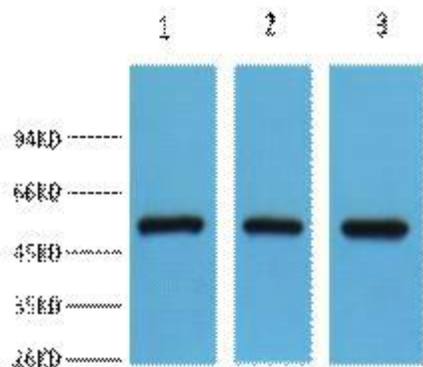
matters needing attention

Avoid repeated freezing and thawing!

Usage suggestions

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

Products Images



Western Blot analysis of various cells using FH Monoclonal Antibody