

FH Monoclonal Antibody(7F1)

Catalog No: YM3073

Reactivity: Human; Mouse; Rat

Applications: WB;IHC;IF

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: Synthetic Peptide of FH

Specificity: The antibody detects endogenous FH proteins.

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

50% Glycerol.

Source: Monoclonal, Mouse

Dilution: WB 1:3000 IF 1:200 IHC 1:50-300

Purification: The antibody was affinity-purified from mouse ascites by affinity-



chromatography using 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 [Isoform Mitochondrial]: Mitochondrion .; [Isoform Cytoplasmic]: Cytoplasm, cytosol . Nucleus . Chromosome . Translocates to the nucleus in response to DNA

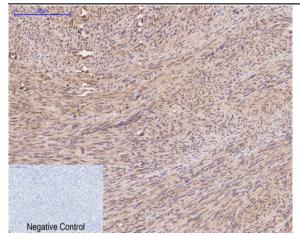
damage: localizes to DNA double-strand breaks (DSBs) following

phosphorylation by PRKDC..

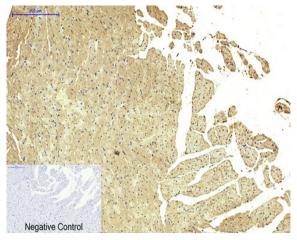
Expression: Expressed in red blood cells; underexpressed in red blood cells (cytoplasm) of

patients with hereditary non-spherocytic hemolytic anemia of unknown etiology.

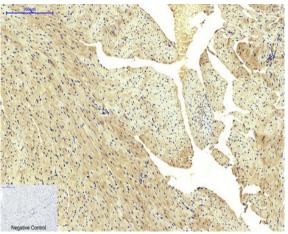
Products Images



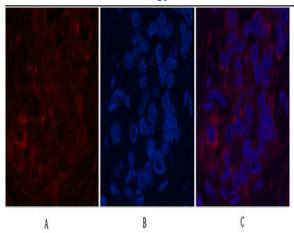
Immunohistochemical analysis of paraffin-embedded Humanuterus tissue. 1,FH Monoclonal Antibody(7F1) was diluted at 1:200(4°C,overnight). 2, Sodium citrate pH 6.0 was used for antibody retrieval(>98°C,20min). 3,Secondary antibody was diluted at 1:200(room tempeRature, 30min). Negative control was used by secondary antibody only.



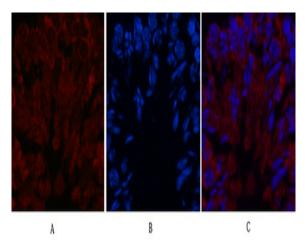
Immunohistochemical analysis of paraffin-embedded Rat-heart tissue. 1,FH Monoclonal Antibody(7F1) was diluted at 1:200(4°C,overnight). 2, Sodium citrate pH 6.0 was used for antibody retrieval(>98°C,20min). 3,Secondary antibody was diluted at 1:200(room tempeRature, 30min). Negative control was used by secondary antibody only.



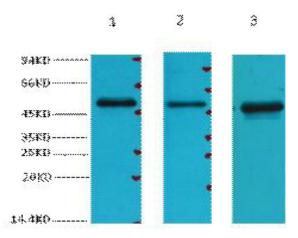
Immunohistochemical analysis of paraffin-embedded Mouseheart tissue. 1,FH Monoclonal Antibody(7F1) was diluted at 1:200(4°C,overnight). 2, Sodium citrate pH 6.0 was used for antibody retrieval(>98°C,20min). 3,Secondary antibody was diluted at 1:200(room tempeRature, 30min). Negative control was used by secondary antibody only.



Immunofluorescence analysis of Human-liver-cancer tissue. 1,FH Monoclonal Antibody(7F1)(red) was diluted at 1:200(4°C,overnight). 2, Cy3 labled Secondary antibody was diluted at 1:300(room temperature, 50min).3, Picture B: DAPI(blue) 10min. Picture A:Target. Picture B: DAPI. Picture C: merge of A+B



Immunofluorescence analysis of Mouse-testis tissue. 1,FH Monoclonal Antibody(7F1)(red) was diluted at 1:200(4°C,overnight). 2, Cy3 labled Secondary antibody was diluted at 1:300(room temperature, 50min).3, Picture B: DAPI(blue) 10min. Picture A:Target. Picture B: DAPI. Picture C: merge of A+B



Western blot analysis of 1) 293T, 2) HepG2, 3) Hela, diluted at 1:3000.