

## **Product Name: FH(7F1)Mouse Monoclonal Antibody**

Catalog #: AMM10956

For research use only.

#### **Summary**

**Description** Mouse monoclonal Antibody

**Host** Mouse

Application WB,IHC,ICC/IF

**Reactivity** Human, Mouse, Rat

ConjugationUnconjugatedModificationUnmodified

**Isotype** IgG

**Clonality** Monoclonal

Form Liquid Concentration 1mg/ml

**Storage** Aliquot and store at -20°C (valid for 12 months). Avoid freeze/thaw cycles.

**Shipping** Ice bags

PBS, pH 7.4, containing 0.5%protective protein, 0.02% New type preservative N as **Buffer** 

Preservative and 50% Glycerol.

**Purification** Affinity purification

## **Application**

**Dilution Ratio** WB 1:1000-1:3000,IHC 1:50-1:300,ICC/IF 1:100-1:200

Molecular Weight 50kDa

## **Antigen Information**

Gene Name FH

**Alternative Names** Fumarate hydratase, mitochondrial (Fumarase) (EC 4.2.1.2)

 Gene ID
 2271.0

 SwissProt ID
 P07954

**Immunogen** Synthetic Peptide of FH

# **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

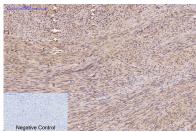


the translation start site used. The N-terminal extended form is targeted to the mitochondrion, where the removal of the extension generates the same form as in the cytoplasm. It is similar to some thermostable class II fumarases and functions as a homotetramer. Mutations in this gene can cause fumarase deficiency and lead to progressive encephalopathy. [provided by RefSeq, Jul 2008],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.,

#### **Research Area**

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

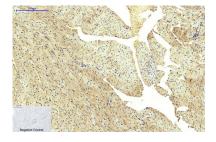
#### **Image Data**



Immunohistochemical analysis of paraffin-embedded Human-uterus 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.



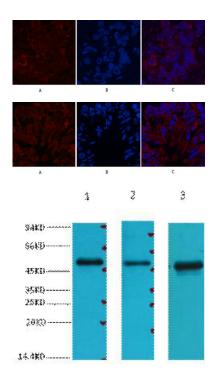
Immunohistochemical analysis of paraffin-embedded Rat-heart tissue. 1,FH Monoclonal Antibody (7F1) was diluted at 1:200 (4°C,overnight) . 2, Sodium citrate pH 6.0 was used for antibody retrieval ( >98°C,20min ) . 3,Secondary antibody was diluted at 1:200 (room tempeRature, 30min) . Negative control was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



Immunohistochemical analysis of paraffin-embedded Mouse-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.

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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.