FH (PT1744R) PT™ Rabbit mAb

CatalogNo: YM9586 Recombinant

Key Features

Host Species
- Rabbit

Reactivity
- Human,Mouse,Rat

Applications
- WB,IHC,IF,IP,ELISA

MW
- 55kD (Calculated)
49kD (Observed)

Isotype
- IgG,Kappa

Storage

Storage*	-15°C to -25°C/1 year(Do not lower than -25°C)
Formulation	PBS, 50% glycerol, 0.05% Proclin 300, 0.05%BSA

Recommended Dilution Ratios

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

Basic Information

Clonality	Monoclonal
Clone Number	PT1744R

Immunogen Information

Specificity	Endogenous

Target Information

Gene name FH

Protein Name Fumarate hydratase, mitochondrial

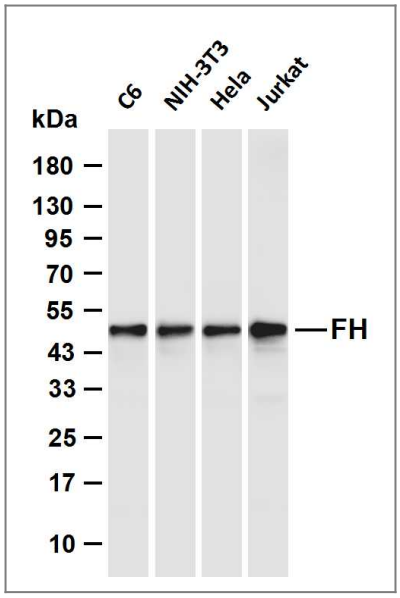
Organism	Gene ID	UniProt ID
Human	2271;	P07954;
Mouse	14194;	P97807;
Rat		P14408;

Cellular Localization [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. .

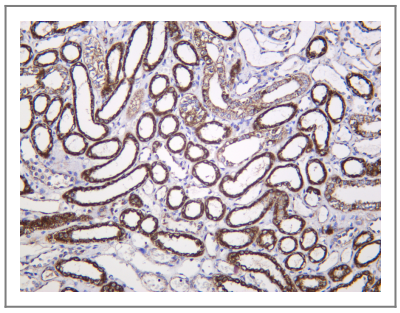
Tissue specificity Expressed in red blood cells; underexpressed in red blood cells (cytoplasm) of patients with hereditary non-spherocytic hemolytic anemia of unknown etiology.

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.,

Validation Data



Various whole cell lysates were separated by 4-20% SDS-PAGE, and the membrane was blotted with anti-FH (PT1744R) 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 (PT1744R) Rabbit antibody

Contact information

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Please scan the QR code to access additional product information:
FH (PT1744R) PT™ Rabbit mAb