

FH/Fumarase Rabbit mAb

Catalog # AP75435

Product Information

Application	WB, FC
Primary Accession	P07954
Reactivity	Rat, Human, Mouse
Host	Rabbit
Clonality	Monoclonal Antibody
Isotype	IgG
Conjugate	Unconjugated
Purification	Affinity Purified
Calculated MW	54637

Additional Information

Gene ID	2271
Other Names	FH
Dilution	WB~~1:1000-1:5000 FC~~1:100-1:200
Format	Liquid in 50mM Tris-Glycine(pH 7.4), 0.15M NaCl, 40%Glycerol, 0.01% sodium azide and 0.05% BSA.
Storage	Store at 4°C short term. Aliquot and store at -20°C long term. Avoid freeze/thaw cycles.

Protein Information

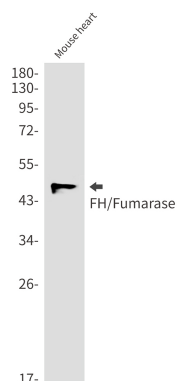
Name	FH {ECO:0000303 PubMed:27037871, ECO:0000312 HGNC:HGNC:3700}
Function	Catalyzes the reversible stereospecific interconversion of fumarate to L-malate (PubMed: 30761759). Experiments in other species have demonstrated that specific isoforms of this protein act in defined pathways and favor one direction over the other (Probable).
Cellular Location	[Isoform Mitochondrial]: Mitochondrion
Tissue Location	Expressed in red blood cells; underexpressed in red blood cells (cytoplasm) of patients with hereditary non-spherocytic hemolytic anemia of unknown etiology.

Background

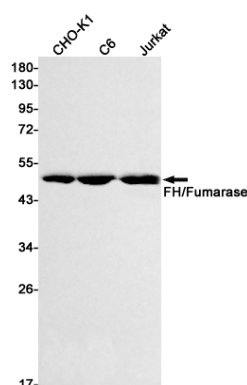
The protein encoded by this gene is an enzymatic component of the tricarboxylic acid (TCA) cycle, or Krebs

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.

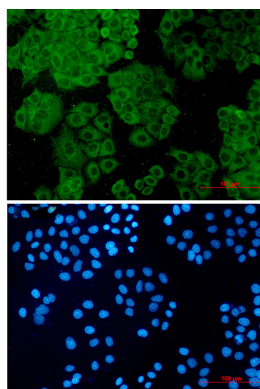
Images



Western blot analysis of FH/Fumarase in mouse heart lysates using FH antibody.



Western blot analysis of FH/Fumarase in CHO-K1, C6, Jurkat lysates using FH/Fumarase antibody.



Immunocytochemistry analysis of FH (green) in HeLa using FH antibody, and DAPI (blue)

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