

FH/Fumarase Rabbit mAb

Catalog # AP75436

Product Information

Application	WB, IHC-P, IHC-F, FC, IP
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~~1:500-1:1000 IHC-P~~N/A IHC-F~~N/A FC~~1:200-1:500 IP~~1:20-1:50
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

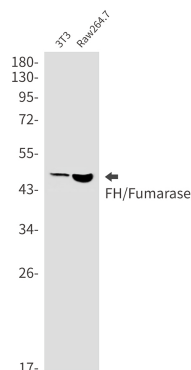
Name	FH {ECO:0000303 PubMed:27037871, ECO:0000312 HGNC:HGNC:3700}
Function	Catalyzes the reversible stereospecific interconversion of fumarate to L-malate (PubMed: 30761759). Experiments in other species have demonstrated that specific isoforms of this protein act in defined pathways and favor one direction 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

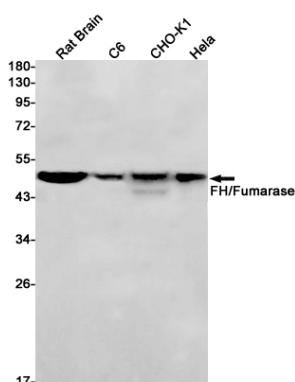
The protein encoded by this gene is an enzymatic component of the tricarboxylic acid (TCA) cycle, or Krebs

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.

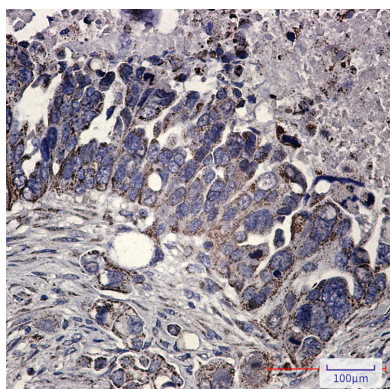
Images



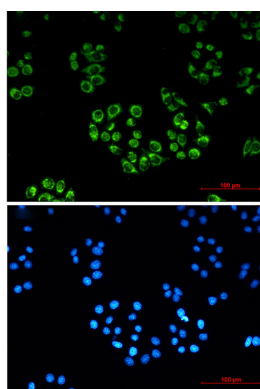
Western blot analysis of FH/Fumarase in 3T3, Raw264.7 lysates using FH antibody.



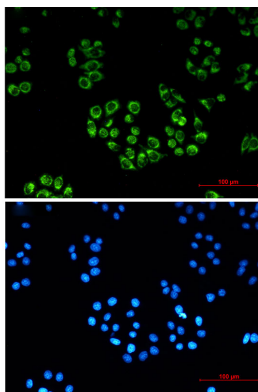
Western blot analysis of FH/Fumarase in rat Brain, C6, CHO-K1, Hela lysates using FH/Fumarase antibody.



Immunohistochemistry analysis of paraffin-embedded Human Cholangiocarcinoma using FH antibody. High-pressure and temperature Sodium Citrate pH 6.0 was used for antigen retrieval.



Immunocytochemistry analysis of FH/Fumarase (green) in Hela using FH/Fumarase antibody, and DAPI (blue).



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