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FH Antibody (N-term)

Affinity Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AW5544

Product Information

Application	WB
Primary Accession	P07954
Reactivity	Human, Mouse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	54637
Isotype	Rabbit IgG
Antigen Source	HUMAN

Additional Information

Gene ID	2271
Antigen Region	107-135
Other Names	Fumarate hydratase, mitochondrial, Fumarase, FH
Dilution	WB~~1:1000
Target/Specificity	This FH antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide between 107-135 amino acids from the N-terminal region of human FH.
Format	Purified polyclonal antibody supplied in PBS with 0.09% (W/V) sodium azide. This antibody is purified through a protein A column, followed by peptide affinity purification.
Storage	Maintain refrigerated at 2-8°C for up to 2 weeks. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.
Precautions	FH Antibody (N-term) is for research use only and not for use in diagnostic or therapeutic procedures.

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.

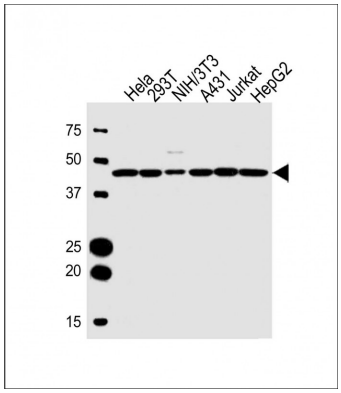
Background

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.

References

Shimada, M., et al. Hum. Genet. 128(4):433-441(2010)
Allegri, G., et al. J. Inherit. Metab. Dis. 33(4):411-419(2010)
Yogev, O., et al. PLoS Biol. 8 (3), E1000328 (2010) :
Yang, Y., et al. Cancer Genet. Cytogenet. 196(1):45-55(2010)
Rikova, K., et al. Cell 131(6):1190-1203(2007)

Images



All lanes : Anti-FH Antibody (N-term) at 1:1000 dilution Lane 1: Hela whole cell lysate Lane 2: 293T whole cell lysate Lane 3: NIH/3T3 whole cell lysate Lane 4: A431 whole cell lysate Lane 5: Jurkat whole cell lysate Lane 6: HepG2 whole cell lysate Lysates/proteins at 20 µg per lane. Secondary Goat Anti-Rabbit IgG, (H+L), Peroxidase conjugated at 1/10000 dilution. Predicted band size : 55 kDa Blocking/Dilution buffer: 5% NFDM/TBST.

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.