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Fumarase / FH, Biotinylated

Peptide-affinity purified goat antibody
Catalog # AF1449b

Product Information

Application	WB, Pep-ELISA
Primary Accession	P07954
Other Accession	NP_000134, 2271, 14194 (mouse), 24368 (rat)
Reactivity	Human
Predicted	Mouse, Rat, Dog
Host	Goat
Clonality	Polyclonal
Isotype	IgG
Calculated MW	54637

Additional Information

Gene ID	2271
Other Names	Fumarate hydratase, mitochondrial, Fumarase, 4.2.1.2, FH
Dilution	WB~~1:1000 Pep-ELISA~~N/A
Format	0.5 mg IgG/ml in Tris saline (20mM Tris pH7.3, 150mM NaCl), 0.02% sodium azide, with 0.5% bovine serum albumin
Storage	Maintain refrigerated at 2-8°C for up to 6 months. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.
Precautions	Fumarase / FH, Biotinylated 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

An approach based on a genome-wide association study reveals candidate loci for narcolepsy. Shimada M, et al. Hum Genet, 2010 Oct. PMID 20677014.
Fumarase: a mitochondrial metabolic enzyme and a cytosolic/nuclear component of the DNA damage response. Yogev O, et al. PLoS Biol, 2010 Mar 9. PMID 20231875.
UOK 262 cell line, fumarate hydratase deficient (FH-/FH-) hereditary leiomyomatosis renal cell carcinoma: in vitro and in vivo model of an aberrant energy metabolic pathway in human cancer. Yang Y, et al. Cancer Genet Cytogenet, 2010 Jan 1. PMID 19963135.
Follow-up examination of linkage and association to chromosome 1q43 in multiple sclerosis. McCauley JL, et al. Genes Immun, 2009 Oct. PMID 19626040.
Novel role of fumarate metabolism in dahl-salt sensitive hypertension. Tian Z, et al. Hypertension, 2009 Aug. PMID 19546378.

Images

Biotinylated EB07874 (0.1 µg/ml) staining of Human Kidney lysate (35 µg protein in RIPA buffer), exactly mirroring its parental non-biotinylated product. Primary incubation was 1 hour. Detected by chemiluminescence, using streptavidin-HRP and using NAP bloc