

ETFDH Rabbit pAb

Catalog # AP98766

Product Information

Application	WB, IHC-P, IHC-F, IF, E
Primary Accession	Q16134
Other Accession	Q16134
Host	Rabbit
Clonality	Polyclonal
Calculated MW	68495
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human ETFDH
Epitope Specificity	41-150/617
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Belongs to the ETF-QO/fixC family. Contains 1 4Fe-4S ferredoxin-type domain.
SUBUNIT	Monomer.
DISEASE	Defects in ETFDH are the cause of glutaric aciduria type 2C (GA2C) [MIM:231680]. GA2C is an autosomal recessively inherited disorder of fatty acid, amino acid, and choline metabolism. It is characterized by multiple acyl-CoA dehydrogenase deficiencies resulting in large excretion not only of glutaric acid, but also of lactic, ethylmalonic, butyric, isobutyric, 2-methyl-butyric, and isovaleric acids.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	Electron-transferring-flavoprotein dehydrogenase in the inner mitochondrial membrane accepts electrons from electron-transfer flavoprotein which is located in the mitochondrial matrix and reduces ubiquinone in the mitochondrial membrane. The protein is synthesized as a 67-kDa precursor which is targeted to mitochondria and processed in a single step to a 64-kDa mature form located in the mitochondrial membrane. Deficiency in electron-transferring-flavoprotein dehydrogenase have been demonstrated in some patients with type II glutaricacidemia. [provided by RefSeq, Jul 2008].

Additional Information

Gene ID	2110
Other Names	ETFQO; MADD; O610010I20Rik; ETFD_BOVIN; ETFDH; ETF-QO; ETF-ubiquinone oxidoreductase; Electron-transferring-flavoprotein dehydrogenase (ETF dehydrogenase); 1.5.5.1; ETFD_HUMAN; ETFD_MOUSE; ETFD_RAT; electron transfer flavoprotein dehydrogenase; electron-transferring-flavoprotein dehydrogenase
Dilution	WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,ICC/IF=1:100-500,IF=1:100-500,ELISA=1:5000-10000

Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
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Protein Information

Name	ETFDH (HGNC:3483)
Function	Links fatty acid beta-oxidation and amino acid catabolism to the respiratory chain by transferring electrons from the electron transfer flavoprotein (ETF) to ubiquinone.
Cellular Location	Mitochondrion inner membrane.

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