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Rabbit Anti-ETFDH antibody

SL13110R

Product Name:	ETFDH
Chinese Name:	电子转移黄素蛋白脱氢酶抗体
Alias:	Electron transfer flavoprotein ubiquinone oxidoreductase; Electron transfer flavoprotein-ubiquinone oxidoreductase; electron transferring flavoprotein dehydrogenase; Electron-transferring-flavoprotein dehydrogenase; ETF dehydrogenase; ETF QO; ETF ubiquinone oxidoreductase; ETF-QO; ETF-ubiquinone oxidoreductase; ETFD_HUMAN; Etfdh; mitochondrial.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Chicken,Dog,Horse,Rabbit,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	65kDa
Cellular localization:	cytoplasmicThe cell membrane
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human ETFDH:41-150/617
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. 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.
PubMed:	PubMed
Product Detail:	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].

Function:
Accepts electrons from ETF and reduces ubiquinone.

Subunit:
Monomer.

Subcellular Location:
Mitochondrion inner membrane.

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.

Similarity:
Belongs to the ETF-QO/fixC family.
Contains 1 4Fe-4S ferredoxin-type domain.

SWISS:
Q16134

Gene ID:
2110

Database links:

Entrez Gene: 768074Cow

Entrez Gene: 2110Human

Entrez Gene: 66841Mouse

Entrez Gene: 295143Rat

Omim: 231675Human

SwissProt: Q2KIG0Cow

SwissProt: Q16134Human

SwissProt: Q921G7Mouse

	SwissProt: Q6UPE1Rat **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.