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ETFA Rabbit pAb

Catalog # AP99215

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	P13804
Other Accession	P13804
Reactivity	Mouse
Predicted	Rat, Pig, Human, Rabbit, Dog, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	35080
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human ETFA
Epitope Specificity	184-260/333
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 alpha-subunit/FixB family.
SUBUNIT	Heterodimer of an alpha and a beta subunit.
Post-translational modifications	The N-terminus is blocked
DISEASE	Defects in ETFA are the cause of glutaric aciduria type 2A (GA2A) [MIM:231680]; also known as glutaricaciduria IIA. GA2A 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	ETFA participates in catalyzing the initial step of the mitochondrial fatty acid beta-oxidation. It shuttles electrons between primary flavoprotein dehydrogenases and the membrane-bound electron transfer flavoprotein ubiquinone oxidoreductase. Defects in electron-transfer-flavoprotein have been implicated in type II glutaricaciduria in which multiple acyl-CoA dehydrogenase deficiencies result in large excretion of glutaric, lactic, ethylmalonic, butyric, isobutyric, 2-methyl-butyric, and isovaleric acids. Two transcript variants encoding different isoforms have been found for this gene. [provided by RefSeq, Jul 2008].

Additional Information

Gene ID	2108
Other Names	EMA; GA2; MADD; 2010200I21Rik; D9Ertd394e; ETF; ETFA_HUMAN; ETFA; Alpha-ETF; ETFA_MOUSE; ETFA_RAT; electron transfer flavoprotein subunit

	alpha; electron-transfer-flavoprotein, alpha polypeptide; glutaric aciduria II; multiple acyl-CoA dehydrogenase deficiency
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
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.

Protein Information

Name	ETFA
Function	Heterodimeric electron transfer flavoprotein that accepts electrons from several mitochondrial dehydrogenases, including acyl-CoA dehydrogenases, glutaryl-CoA and sarcosine dehydrogenase (PubMed:10356313, PubMed:15159392, PubMed:15975918, PubMed:27499296, PubMed:9334218). It transfers the electrons to the main mitochondrial respiratory chain via ETF-ubiquinone oxidoreductase (ETF dehydrogenase) (PubMed:9334218). Required for normal mitochondrial fatty acid oxidation and normal amino acid metabolism (PubMed:12815589, PubMed:1430199, PubMed:1882842).
Cellular Location	Mitochondrion matrix

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