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Rabbit Anti-ETFA antibody

SL0494R

Product Name:	ETFA
Chinese Name:	电子转移黄素蛋白α抗体
Alias:	ETF-alpha; Electron transfer flavoprotein subunit alpha; electron-transfer-flavoprotein, alpha polypeptide; mitochondrial; Alpha ETF; Alpha-ETF; Electron transfer flavoprotein alpha polypeptide; Electron transfer flavoprotein alpha subunit; Electron transfer flavoprotein subunit alpha; Electron transfer flavoprotein subunit alpha mitochondrial; Electron transfer flavoprotein subunit alpha, mitochondrial; Electron transferring flavoprotein alpha polypeptide; EMA; ETFA; ETFA_HUMAN; GA2.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Pig,Cow,Horse,Rabbit,Sheep,Xenopuslaevis
Applications:	ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:50-200（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	37kDa
Cellular localization:	cytoplasmicMitochondrion
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human ETFA:184-260/333
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:	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].

Function:
The electron transfer flavoprotein serves as a specific electron acceptor for several dehydrogenases, including five acyl-CoA dehydrogenases, glutaryl-CoA and sarcosine dehydrogenase. It transfers the electrons to the main mitochondrial respiratory chain via ETF-ubiquinone oxidoreductase (ETF dehydrogenase).

Subunit:
Heterodimer of an alpha and a beta subunit.

Subcellular Location:
Mitochondrion matrix.

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.

Similarity:
Belongs to the ETF alpha-subunit/FixB family.

SWISS:
P13804

Gene ID:
2108

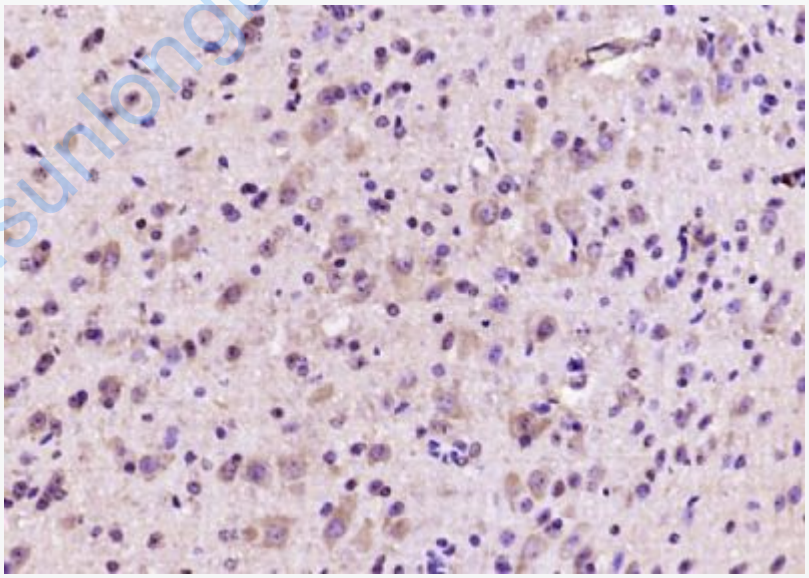
Database links:

Entrez Gene: 2108Human

Entrez Gene: 110842Mouse

Entrez Gene: 300726Rat

Omim: 608053Human

	SwissProt: P13804Human SwissProt: Q99LC5Mouse SwissProt: P13803Rat Unigene: 39925Human Unigene: 290853Mouse Unigene: 32496Rat **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications. Involvement in disease:Defects in ETFA are the cause of glutaric aciduria type 2A (GA2A); 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.
Picture:	 Paraformaldehyde-fixed, paraffin embedded (mouse brain); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (ETFA) Polyclonal Antibody, Unconjugated

	(SL0494R) at 1:200 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.