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Rabbit Anti-ETHE1 antibody

SL7610R

Product Name:	ETHE1
Chinese Name:	乙基丙二酸脑病蛋白抗体
Alias:	mitochondrial; Ethe1; ETHE1 protein, mitochondrial precursor; ETHE1_HUMAN; ethylmalonic encephalopathy 1; Ethylmalonic encephalopathy protein 1; hepatoma subtracted clone one; Hepatoma subtracted clone one protein; HSCO; Protein ETHE1; YF13H12.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Dog,Pig,Cow,Rabbit,Sheep,
Applications:	ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800IF=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:	26kDa
Cellular localization:	The nucleuscytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human ETHE1:191-254/254
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:	Probably plays an important role in metabolic homeostasis in mitochondria. May function as a nuclear-cytoplasmic shuttling protein that binds transcription factor RELA/NFKB3 in the nucleus and exports it to the cytoplasm. Suppresses p53-induced apoptosis by preventing nuclear localization of RELA.

Involvement in disease:Defects in ETHE1 are a cause of ethylmalonic encephalopathy (EE) . EE is an autosomal recessive disorder characterized by neurodevelopmental delay and regression, recurrent petechiae, acrocyanosis, diarrhea, leading to death in the first decade of life. It is also associated with persistent lactic acidemia and ethylmalonic and methylsuccinic aciduria.

Function:
Probably plays an important role in metabolic homeostasis in mitochondria. May function as a nuclear-cytoplasmic shuttling protein that binds transcription factor RELA/NFKB3 in the nucleus and exports it to the cytoplasm. Suppresses p53-induced apoptosis by preventing nuclear localization of RELA.

Subunit:
Interacts with RELA.

Subcellular Location:
Cytoplasm. Nucleus. Mitochondrion matrix. Note=According to PubMed:12398897, it is cytoplasmic and nuclear. According to PubMed:14732903, it is found in the mitochondrial matrix.

Tissue Specificity:
Ubiquitously expressed.

DISEASE:
Defects in ETHE1 are a cause of ethylmalonic encephalopathy (EE) [MIM:602473]. EE is an autosomal recessive disorder characterized by neurodevelopmental delay and regression, recurrent petechiae, acrocyanosis, diarrhea, leading to death in the first decade of life. It is also associated with persistent lactic acidemia and ethylmalonic and methylsuccinic aciduria.

Similarity:
Belongs to the metallo-beta-lactamase superfamily. Glyoxalase II family.

SWISS:
O95571

Gene ID:
23474

Database links:

Entrez Gene: 23474Human

Entrez Gene: 66071Mouse

Entrez Gene: 292710Rat

	Omim: 608451Human SwissProt: O95571Human SwissProt: Q9DCM0Mouse Unigene: 7486Human Unigene: 29553Mouse Unigene: 14691Rat **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.