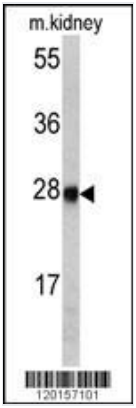
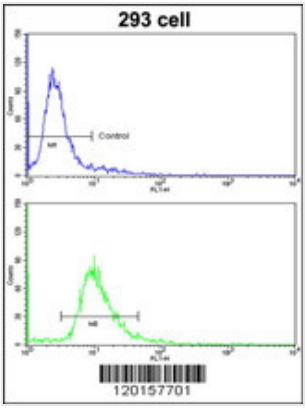
ETHE1 Antibody – Rabbit Polyclonal

PRODUCT OVERVIEW

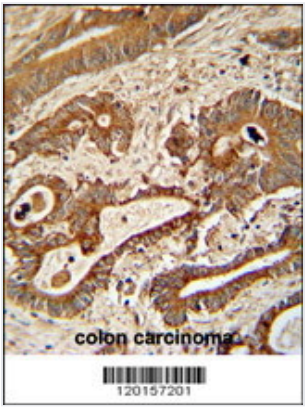
Product Name	ETHE1, Polyclonal Antibody
Full Product Name	ETHE1, ID (Protein ETHE1, Mitochondrial, Ethylmalonic Encephalopathy Protein 1, Hepatoma Subtracted Clone One Protein, HSCO) (FITC)
Product Gene Name	anti-ETHE1 antibody
Clonality	Polyclonal
Isotype	IgG
Host	Rabbit
Species Reactivity	Human, Mouse
Form/Format	Supplied as a liquid in PBS, pH 7.2. No preservative added. Labeled with Fluorescein Isothiocyanate (FITC).
Purity/Purification	Purified by Ammonium Sulfate Precipitation.
Specificity	Recognizes human ETHE1. Species Crossreactivity: mouse.
Preparation and Storage	Store product at 4 degree C if to be used immediately within two weeks. For long-term storage, aliquot to avoid repeated freezing and thawing and store at -20 degree C. Aliquots are stable at -20 degree C for 12 months after receipt. Dilute required amount only prior to immediate use. Further dilutions can be made in assay buffer.
Molecular Weight	29.1 kDa (267aa) confirmed by MALDI-TOF
OMIM	602473
3D Structure	ModBase 3D Structure for O95571
Immunogen	Synthetic peptide selected from the center region of human ETHE1 (KLH).
Conjugate	FITC
Note	Preservative Free
Product Categories/Family for anti-ETHE1 antibody	Antibodies; Enzymes
Applications Tested/Suitable for anti-ETHE1 antibody	Western Blot (WB), ELISA (EIA)
Application Notes for anti-ETHE1 antibody	WB: 1:50-1:100 ELISA: 1:1,000 Applications are based on unconjugated antibody.
Related Product Information for anti-ETHE1 antibody	MBS6475047 is an antibody targeting ETHE1. Also known as ETHE1, ID (Protein ETHE1, Mitochondrial, Ethylmalonic Encephalopathy Protein 1, Hepatoma Subtracted Clone One Protein, HSCO) (FITC). Reactive in Human, Mouse. Host: Rabbit.
Product References and Citations for anti-ETHE1 antibody	1. Tiranti, V., Nat. Med. 15(2), 200-205 (2009). 2. Mineri, R., J. Med. Genet. 45(7), 473-478 (2008).
Other Notes	Small volumes of anti-ETHE1 antibody vial(s) may occasionally become entrapped in the seal of the product vial during shipment and storage. If necessary, briefly centrifuge the vial on a tabletop centrifuge to dislodge any liquid in the container`s cap. Certain products may require to ship with dry ice and additional dry ice fee may apply.



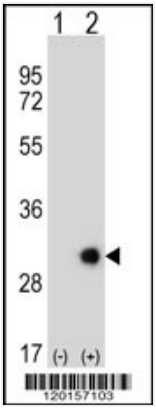
Western Blot (WB) of anti-ETHE1 antibody
Western blot analysis of ETHE1 antibody (Center) in mouse kidney tissue lysates (35ug/lane). ETHE1 (arrow) was detected using the purified Pab.



Flow Cytometry (FC/FACS) of anti-ETHE1 antibody
Flow cytometric analysis of 293 cells using ETHE1 Antibody (Center)(bottom histogram) compared to a negative control cell (top histogram). FITC-conjugated goat-anti-rabbit secondary antibodies were used for the analysis.



Immunohistochemistry (IHC) of anti-ETHE1 antibody
ETHE1 Antibody (Center) immunohistochemistry analysis in formalin fixed and paraffin embedded human colon carcinoma followed by peroxidase conjugation of the secondary antibody and DAB staining. This data demonstrates the use of the ETHE1 Antibody (Center) for immunohistochemistry. Clinical relevance has not been evaluated.



Western Blot (WB) of anti-ETHE1 antibody
Western blot analysis of ETHE1 (arrow) using rabbit polyclonal ETHE1 Antibody (Center). 293 cell lysates (2 ug/lane) either nontransfected (Lane 1) or transiently transfected (Lane 2) with the ETHE1 gene.

ADDITIONAL DETAILS

NCBI/Uniprot data below describe general gene information for ETHE1. It may not necessarily be applicable to this product.

NCBI GI #	41327741
NCBI GeneID	23474
NCBI Accession #	NP_055112
UniProt Primary Accession #	O95571
UniProt Secondary Accession #	Q96HR0; Q9H001
UniProt Related Accession #	O95571
GenBank Nucleotide #	NM_014297.4
NCBI Official Full Name	persulfide dioxygenase ETHE1, mitochondrial isoform 1
NCBI Official Synonym Full Names	ETHE1, persulfide dioxygenase
NCBI Official Symbol	ETHE1
NCBI Official Synonym Symbols	HSCO; YF13H12
UniProt Protein Name	Persulfide dioxygenase ETHE1, mitochondrial
UniProt Synonym Protein Names	Ethylmalonic encephalopathy protein 1; Hepatoma subtracted clone one protein; Sulfur dioxygenase ETHE1

Protein Family	Persulfide dioxygenase
UniProt Gene Name	ETHE1
UniProt Synonym Gene Names	HSCO
UniProt Entry Name	ETHE1_HUMAN
NCBI Protein Information	persulfide dioxygenase ETHE1, mitochondrial
NCBI Summary for ETHE1	This gene encodes a member of the metallo beta-lactamase family of iron-containing proteins involved in the mitochondrial sulfide oxidation pathway. The encoded protein catalyzes the oxidation of a persulfide substrate to sulfite. Certain mutations in this gene cause ethylmalonic encephalopathy, an infantile metabolic disorder affecting the brain, gastrointestinal tract and peripheral vessels. Alternative splicing results in multiple transcript variants encoding different isoforms. [provided by RefSeq, Mar 2016]
UniProt Comments for ETHE1	ETHE1: 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. 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. Belongs to the metallo-beta-lactamase superfamily. Glyoxalase II family. Protein type: Hydrolase; Mitochondrial; EC 1.13.11.18 Chromosomal Location of Human Ortholog: 19q13.31 Cellular Component: nucleoplasm; mitochondrion; mitochondrial matrix; cytoplasm Molecular Function: iron ion binding; sulfur dioxygenase activity Biological Process: glutathione metabolic process; sulfur amino acid metabolic process; sulfur amino acid catabolic process Disease: Encephalopathy, Ethylmalonic
Research Articles on ETHE1	**1.** observations indicate the severe impact of ETHE1 deficiency on cellular physiology and redox state
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