

## ETHE1 Human

**Description:**ETHE1 Human Recombinant produced in E.coli is a single, non-glycosylated polypeptide chain containing 267 amino acids (13-254) and having a molecular mass of 29.1kDa.ETHE1 is fused to a 25 amino acid His-tag at N-terminus & purified by proprietary chromatographic techniques.

**Catalog #:**PRPS-1034

For research use only.

**Synonyms:**Ethylmalonic encephalopathy protein 1, HSCO, Hepatoma subtracted clone one protein, YF13H12, protein ETHE1 mitochondrial, D83198, EC 3.1.2.6.

**Source:**E.coli.

**Physical Appearance:**Sterile Filtered colorless solution.

**Amino Acid Sequence:**MGSSHHHHHH SSGLVPRGSH MGSMLSQRG GSGAPILLRQ  
MFEPVSTFT YLLGDRESRE AVLIDPVLET APRDAQLIKE LGLRLLYAVN THCHADHITG  
SGLLRSLLPG CQSVISRLSG AQADLHIEDG DSIRFGRFAL ETRASPGHTP GCVTFVLNDH  
SMAFTGDALL IRGCGRTDFQ QGCAKTYHS VHEKIFTLPG DCLIYPAHDY HGFTVSTVEE  
ERTLNPRLL SC

**Purity:**Greater than 90% as determined by SDS-PAGE.

### Formulation:

The ETHE1 solution (1mg/ml) contains 20mM Tris-HCl buffer (pH 8.0), 100mM NaCl and 10% glycerol.

### Stability:

Store at 4°C if entire vial will be used within 2-4 weeks. Store, frozen at -20°C for longer periods of time. For long term storage it is recommended to add a carrier protein (0.1% HSA or BSA). Avoid multiple freeze-thaw cycles.

### Usage:

NeoBiolab's products are furnished for LABORATORY RESEARCH USE ONLY. The product may not be used as drugs, agricultural or pesticidal products, food additives or household chemicals.

### Introduction:

ETHE1 is a mitochondrial sulfur dioxygenase involved in catabolism of sulfide that accumulates to toxic levels in ethylmalonic encephalopathy. Mutations of ETHE1 were detected in all the typical ethylmalonic encephalopathy patients analysed, but no ETHE1 mutations were identified in patients presenting with early onset progressive encephalopathy with ethylmalonic aciduria.

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