

Recombinant human ETHE1 protein

Catalog Number: ATGP1382

PRODUCT INFORMATION

Expression system

E.coli

Domain

13-254aa

UniProt No.

O95571

NCBI Accession No.

NP_055112

Alternative Names

Protein ETHE1 mitochondrial, Protein ETHE1, mitochondrial,

PRODUCT SPECIFICATION

Molecular Weight

29.1 kDa (267aa) confirmed by MALDI-TOF

Concentration

1mg/ml (determined by Bradford assay)

Formulation

Liquid in. 20mM Tris-HCl buffer (pH 8.0) containing 10% glycerol, 0.1M NaCl

Purity

> 90% by SDS-PAGE

Tag

His-Tag

Application

SDS-PAGE

Storage Condition

Can be stored at +2C to +8C for 1 week. For long term storage, aliquot and store at -20C to -80C. Avoid repeated freezing and thawing cycles.

BACKGROUND

Description

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. Recombinant human ETHE1 protein, fused to His-tag at N-terminus, was expressed in E. coli and purified by using conventional chromatography.

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Amino acid Sequence

<MGSSHHHHHH SSGLVPRGSH MGSIM>LSQRG GSGAPILLRQ MFEPVSCTFT YLLGDRESRE AVLIDPVLET
APRDAQLIKE LGLRLLYAVN THCHADHITG SGLLRSLPG CQSVISRLSG AQADLHIEDG DSIRFGRFAL ETRASPGHTP
GCVTFVLNDH SMAFTGDALL IRGCGRTDFQ QGCAKTLYHS VHEKIFTLPG DCLIYPAHDY HGFTVSTVEE ERTLNPRRTL
SCEEFVKIMG NLNLPKPQI DFAVPANMRC GVQTPTA

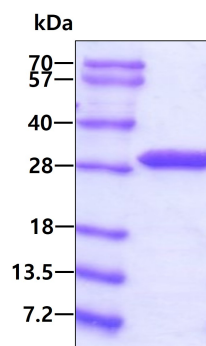
General References

Tiranti V. et al. (2009) Nat Med. 15:200-205.

Mineri R. et al. (2008) J Med Genet. 45:473-478

DATA

SDS-PAGE



3ug by SDS-PAGE under reducing condition and visualized by coomassie blue stain.