

Product information

ETHE1, 13-254aa

Human, His-tagged, Recombinant, *E.coli*

Cat. No. IBATGP1382

Full name: Protein ETHE1, mitochondrial

NCBI Accession No.: NP_055112

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.

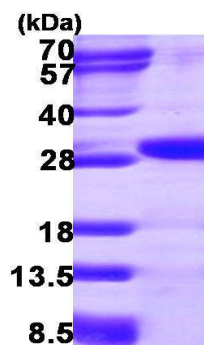
Form: Liquid. 20mM Tris-HCl buffer (pH8.0) containing 10% glycerol

0.1M NaCl

Molecular Weight: 29.1 kDa (267aa), confirmed by MALDI-TOF

Purity: > 90% by SDS - PAGE

Concentration: 1.0 mg/ml (determined by Bradford assay)



12% SDS-PAGE (3ug)

Sequences of amino acids:

MGSSHHHHH SSGLVPRGSH MGSMLSQRG GSGAPILLRQ MFEPVSCFT YLLGDRESRE AVLIDPVLET APRDAQLIKE LGLRLLYAVN
THCHADHITG SGLLRSLLPQ CQSVISRLSG AQADLHIEDG DSIRFGRFAL ETRASPGHTP GCVTFVLNDH SMAFTGDALL IRGCGRTDFQ
QGCAKTYHS VHEKIFTLPG DCLLYPAHDY HGFTVSTVEE ERTLNPRLTL SCEEFVKIMG NLNLPKPQQI 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

Storage: Can be stored at +4°C short term (1-2 weeks). For long term storage, aliquot and store at -20°C or -70°C.

Avoid repeated freezing and thawing cycles.

For research use only. This product is not intended or approved for human, diagnostics or veterinary use.