Human Fumarate Hydratase / FH Protein (His Tag)

Catalog Number: 12115-H08E

General Information

Gene Name Synonym:

FMRD; HLRCC; LRCC; MCL; MCUL1

Protein Construction:

A DNA sequence encoding the mature form of human FH (P07954-1) (Ala 45-Lys 510) was fused with a polyhistidine tag at the C-terminus and an initial Met at the N-terminus.

Source: Human

Expression Host: E. coli

QC Testing

Purity: > 85 % as determined by SDS-PAGE

Bio Activity:

Measured by its ability to transform 1umole of Fumarate to L-malate per minute at pH 7.5 at 37° C. Specific activity is > 25 unit/mg.

Endotoxin:

Please contact us for more information.

Stability:

Samples are stable for up to twelve months from date of receipt at -70 ℃

Predicted N terminal: Met

Molecular Mass:

The recombinant human FH consisting of 477 amino acids and has a calculated molecular mass of 52 kDa. It migrates as an approximately 45 kDa band in SDS-PAGE under reducing conditions.

Formulation:

Lyophilized from sterile 10mM Tris, 5mM EDTA, 1mM DTT, pH 7.5

Normally 5 % - 8 % trehalose, mannitol and 0.01% Tween80 are added as protectants before lyophilization. Specific concentrations are included in the hardcopy of COA. Please contact us for any concerns or special requirements.

Usage Guide

Storage:

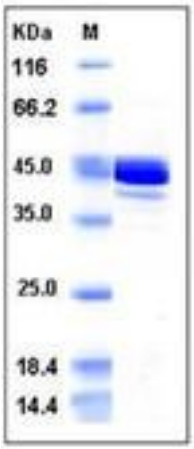
Store it under sterile conditions at -20℃ to -80℃ upon receiving. Recommend to aliquot the protein into smaller quantities for optimal storage.

Avoid repeated freeze-thaw cycles.

Reconstitution:

Detailed reconstitution instructions are sent along with the products.

SDS-PAGE:



Protein Description

Fumarate Hydratase (FH) is an enzymatic component of the tricarboxylic acid (TCA) cycle, or Krebs cycle, and catalyzes the formation of L-malate from fumarate. It exists in both a cytosolic form and an N-terminal extended form, differing only in the translation start site used. The N-terminal extended form is targeted to the mitochondrion, where the removal of the extension generates the same form as in the cytoplasm. Fumarate Hydratase is similar to some thermostable class II fumarases and functions as a homotetramer. Mutations in this gene can cause fumarase deficiency and lead to progressive encephalopathy. Individuals with hemizygous germline fumarate hydratase (FH) mutations are predisposed to renal cancer. These tumors predominantly exhibit functional inactivation of the remaining wild-type allele, implicating FH inactivation as a tumor-promoting event.

References

1.King A, *et al.* (2005) Succinate dehydrogenase and fumarate hydratase: linking mitochondrial dysfunction and cancer. Oncogene. 25(34): 4675-82. 2.Alam NA, *et al.* (2003) Genetic and functional analyses of FH mutations in multiple cutaneous and uterine leiomyomatosis, hereditary leiomyomatosis and renal cancer, and fumarate hydratase deficiency. Hum Mol Genet.12(11): 1241-52. 3.Pollard PJ, *et al.* (2003) The TCA cycle and tumorigenesis: the examples of fumarate hydratase and succinate dehydrogenase. Ann Med. 35(8): 632-9.