

# Recombinant human Fumarase/FH protein

Catalog Number: FMR0905

## PRODUCT INFORMATION

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### Expression system

E.coli

### Domain

44-510aa

### UniProt No.

P07954

### NCBI Accession No.

NP\_000134

### Alternative Names

Fumarate hydratase, FH, HLRCC, LRCC, MCL, MCuL1, Fumarate hydratase Fumarase, Fumarate hydratase mitochondrial, MCuL 1, Multiple hereditary cutaneous leiomyomata.

## PRODUCT SPECIFICATION

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### Molecular Weight

50.2 kDa (467aa) confirmed by MALDI-TOF

### Concentration

1mg/ml (determined by Bradford assay)

### Formulation

Liquid in. 20mM Tris-HCl buffer (pH 8.0)

### Purity

> 95% by SDS-PAGE

### Endotoxin level

< 1 EU per 1ug of protein (determined by LAL method)

### Biological Activity

Specific activity is > 25 unit/mg, and is defined as the amount of enzyme that cleaves 1umole of L-Malate to Fumarate per minute at pH 7.5 at 37C.

### Tag

Non-Tagged

### Application

SDS-PAGE, Enzyme Activity

### 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.

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## BACKGROUND

### Description

Fumarase (Fumarate hydratase) is an enzyme that catalyzes the reversible hydration/dehydration of fumarate to S-malate and is involved in the tricarboxylic acid (TCA), or Krebs cycle. This enzyme exists in both a cytosolic form and an N-terminal extended mitochondrial form. The N-terminal extended form is targeted to the mitochondrion, where the removal of the extension is the same form as in the cytoplasm. Fumarase deficiency can lead to progressive encephalopathy, cerebral atrophy and developmental delay and this enzyme also is thought to act as a tumor suppressor. Recombinant Fumarase was expressed in *E. coli* and was purified by conventional chromatography techniques.

### Amino acid Sequence

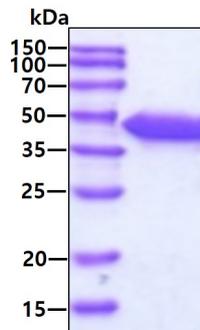
MASQNSFRIE YDTFGELKVP NDKYYGAQTV RSTMNFKIGG VTERMPTPVI KAFGILKRAA AEVNQDYGLD PKIANAIMKA ADEVAEGKLN DHFPLVWWT GSGTQTNMNV NEVISNRAIE MLGGELGSKI PVHPNDHVNK SQSSNDTFPT AMHIAAAIEV HEVLLPGLQK LHDALDAKSK EFAQIIGR THTQDAVPLT LGQEFSGYVQ QVKYAMTRIK AAMPRIYELA AGGTAVGTGL NTRIGFAEKV AAKVAALTGL PFVTAPNKFE ALAAHDALVE LSGAMNTTAC SLMKIANDIR FLGSGPRSGL GELILPENEP GSSIMPGKVN PTQCEAMTMV AAQVMGNHVA VTVGGSNHGF ELNVFKPMMI KNLHLSARLL GDASVSFTEN CVVGIQANTE RINKLMNESL MLVTALNPHI GYDKAAKIAK TAHKNGSTLK ETAIELGYLT AEQFDEWVKP KDMLGPK

### General References

Lehtonen R., et al. (2003) *J. Med. Genet.* 40 (3): e19.  
Toro JR., et al. (2003) *Am. J. Hum. Genet.* 73(1): 95-106.

## DATA

### SDS-PAGE



3 $\mu$ g by SDS-PAGE under reducing condition and visualized by coomassie blue stain.