abcam

Product datasheet

Recombinant human FH protein ab168013

1 Image

Overview

Product name	Recombinant human FH protein
Protein length	Full length protein

Description

Nature	Recombinant
Source	Escherichia coli
Amino Acid Sequence	
Accession	P07954
Species	Human
Sequence	MASQNSFRIE YDTFGELKVP NDKYYGAQTV RSTMNFKIGG VTERMPTPVI KAFGILKRAA AEVNQDYGLD PKIANAIMKA ADEVAEGKLN DHFPLVVWQT GSGTQTNMNV NEVISNRAIE MLGGELGSKI PVHPNDHVNK SQSSNDTFPT AMHIAAAIEV HEVLLPGLQK LHDALDAKSK EFAQIIKIGR THTQDAVPLT LGQEFSGYVQ QVKYAMTRIK AAMPRIYELA AGGTAVGTGL NTRIGFAEKV AAKVAALTGL PFVTAPNKFE ALAAHDALVE LSGAMNTTAC SLMKIANDIR FLGSGPRSGL GELILPENEP GSSIMPGKVN PTQCEAMTMV AAQVMGNHVA VTVGGSNGHF ELNVFKPMMI KNVLHSARLL GDASVSFTEN CVVGIQANTE RINKLMNESL MLVTALNPHI GYDKAAKIAK TAHKNGSTLK ETAIELGYLT AEQFDEWVKP KDMLGPK
Molecular weight	50 kDa
Amino acids	44 to 510

Specifications

Our Abpromise guarantee covers the use of **ab168013** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Biological activity	Specific activity is > 1.0 unit/mg, and is defined as the amount of enzyme that cleaves 1 µmole of L-Malate to Fumarate per minute at pH 7.5 at 25°C
Applications	SDS-PAGE Functional Studies
Endotoxin level	< 1.000 Eu/µg
Purity	>95% by SDS-PAGE .
Form	Liquid
Additional notes	Centrifuge the vial prior to opening

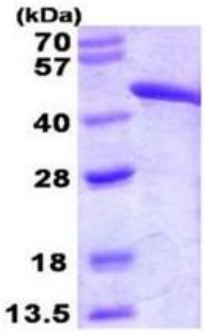
Preparation and Storage

Stability and Storage	Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C or -80°C. Avoid freeze / thaw cycle. pH: 8.00 Constituent: 0.32% Tris HCl This product is an active protein and may elicit a biological response in vivo, handle with caution.

General Info

Function	Also acts as a tumor suppressor.
Pathway	Carbohydrate metabolism; tricarboxylic acid cycle; (S)-malate from fumarate: step 1/1.
Involvement in disease	Defects in FH are the cause of fumarase deficiency (FHD) [MIM:606812]; also known as fumaricaciduria. FHD is characterized by progressive encephalopathy, developmental delay, hypotonia, cerebral atrophy and lactic and pyruvic acidemia. Defects in FH are the cause of multiple cutaneous and uterine leiomyomata (MCUL1) [MIM:150800]. MCUL1 is an autosomal dominant condition in which affected individuals develop benign smooth muscle tumors (leiomyomata) of the skin. Affected females also usually develop leiomyomata of the uterus (fibroids). Defects in FH are the cause of hereditary leiomyomatosis and renal cell cancer (HLRCC) [MIM:605839].
Sequence similarities	Belongs to the class-II fumarase/aspartase family. Fumarase subfamily.
Cellular localization	Cytoplasm and Mitochondrion.

Images



SDS-PAGE - Recombinant human FH protein (ab168013)

15% SDS-PAGE analysis of 3µg ab168013.

Please note: All products are "FOR RESEARCH USE ONLY AND ARE NOT INTENDED FOR DIAGNOSTIC OR THERAPEUTIC USE"

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