

Recombinant Human FH/Fumarase protein ab82790

1 Image

Description	
Product name	Recombinant Human FH/Fumarase protein
Purity	> 95 % SDS-PAGE.
Expression system	Escherichia coli
Protein length	Full length protein
Animal free	No
Nature	Recombinant
Species	Human
Sequence	MASQNSFRIE YDTFGELKVP NDKYYGAQTV RSTMNFKIGG VTERMPTPVI KAFGILKRAA AEVNQDYGLD PKIANAIMKA ADEVAEGKLN DHFPLVWQQT 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 KNLHSARLL GDASVSFTEN CVVGIQANTE RINKLMNESL MLVTALNPHI GYDKAAKIAK TAHKNGSTLK ETAIELGYLT AEQFDEWVKP KDMLGPK

Specifications	
Our Abpromise guarantee covers the use of ab82790 in the following tested applications.	
The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.	
Applications	SDS-PAGE
Form	Liquid
Additional notes	Endotoxin Level: < 1.0 EU per 1µg of protein (determined by LAL method) This product was previously labelled as FH

Preparation and Storage

Stability and Storage

Shipped at 4°C. Upon delivery aliquot and store at -20°C or -80°C. Avoid repeated freeze / thaw cycles.

pH: 8.00

Constituent: 0.316% Tris HCl

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].

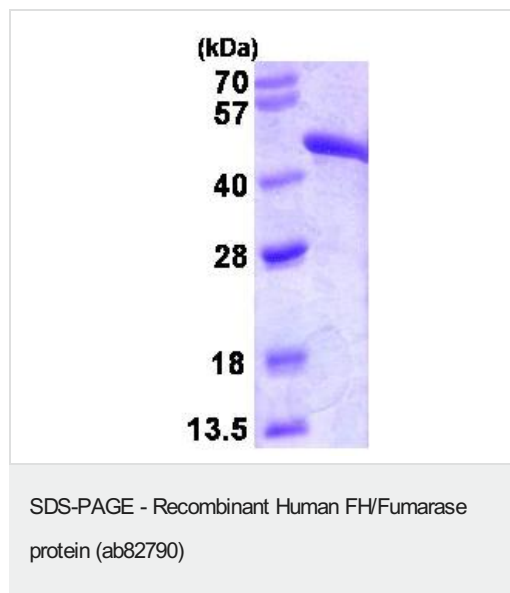
Sequence similarities

Belongs to the class-II fumarase/aspartase family. Fumarase subfamily.

Cellular localization

Cytoplasm and Mitochondrion.

Images



ab82790 on 15% SDS-PAGE (3µg)

Please note: All products are "FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC PROCEDURES"

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- Response to your inquiry within 24 hours
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- Extensive multi-media technical resources to help you
- We investigate all quality concerns to ensure our products perform to the highest standards

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