abcam

Product datasheet

Recombinant Human SDHA protein (Tagged) ab226268

3 Images

Description

Product name	Recombinant Human SDHA protein (Tagged)
Purity	> 90 % SDS-PAGE.
Expression system	Escherichia coli
Accession	P31040
Protein length	Protein fragment
Animal free	No
Nature	Recombinant
Species	Human
Sequence	SAKVSDSISAQYPVVDHEFDAVVVGAGGAGLRAAFGLSEAGFNTACVTKL FPTRSHTVAAQGGINAALGNMEEDNWRWHFYDTVKGSDWLGDQDAIHYMT EQAPAAVVELENYGMPFSRTEDGKIYQRAFGGQSLKFGKGGQAHRCCCVA DRTGHSLLHTLYGRSLRYDTSYFVEYFALDLLMENGECRGVIALCIEDGS IHRIRAKNTVVATGGYGRTYFSCTSAHTSTGDGTAMITRAGLPCQDLEFV
Predicted molecular weight	54 kDa including tags
Amino acids	44 to 293
Tags	GST tag N-Terminus

Specifications

Our **Abpromise guarantee** covers the use of **ab226268** 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 Mass Spectrometry
Mass spectrometry	LC-MS/MS
Form	Liquid

Preparation and Storage

Stability and Storage

Shipped at 4°C. Upon delivery aliquot. Store at -20°C or -80°C. Avoid freeze / thaw cycle.

Constituents: 50% Glycerol, Tris buffer

General Info

Function

Flavoprotein (FP) subunit of succinate dehydrogenase (SDH) that is involved in complex II of the mitochondrial electron transport chain and is responsible for transferring electrons from succinate to ubiquinone (coenzyme Q).

Pathway

Carbohydrate metabolism; tricarboxylic acid cycle; fumarate from succinate (eukaryal route): step 1/1.

Involvement in disease

Defects in SDHA are a cause of mitochondrial complex II deficiency (MT-C2D) [MIM:252011]. A disorder of the mitochondrial respiratory chain with heterogeneous clinical manifestations. Clinical features include psychomotor regression in infants, poor growth with lack of speech development, severe spastic quadriplegia, dystonia, progressive leukoencephalopathy, muscle weakness, exercise intolerance, cardiomyopathy. Some patients manifest Leigh syndrome or Kearns-Sayre syndrome.
Defects in SDHA are a cause of Leigh syndrome (LS) [MIM:256000]. LS is a severe disorder characterized by bilaterally symmetrical necrotic lesions in subcortical brain regions.
Defects in SDHA are the cause of cardiomyopathy dilated type 1GG (CMD1GG) [MIM:613642]. CMD1GG is a disorder characterized by ventricular dilation and impaired systolic function, resulting in congestive heart failure and arrhythmia. Patients are at risk of premature death.

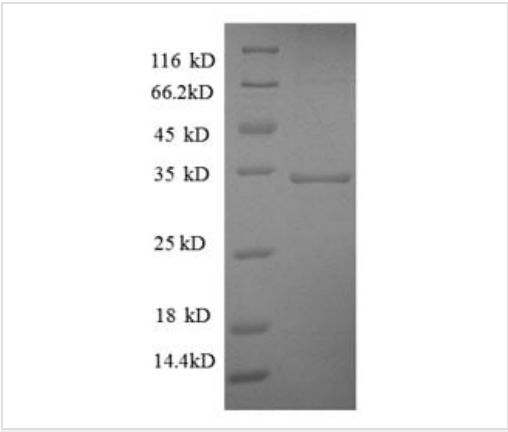
Sequence similarities

Belongs to the FAD-dependent oxidoreductase 2 family. FRD/SDH subfamily.

Cellular localization

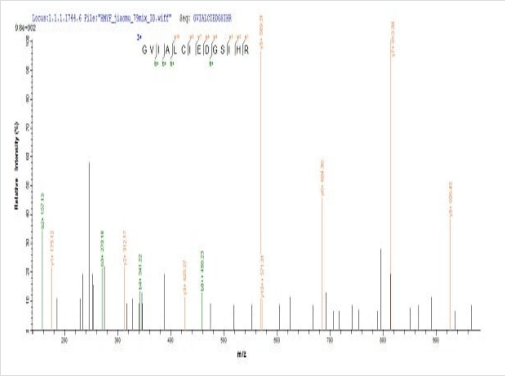
Mitochondrion inner membrane.

Images



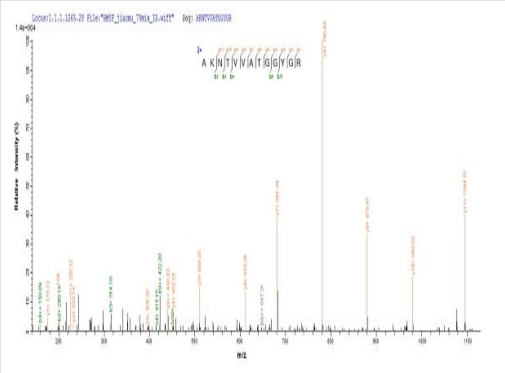
SDS-PAGE - Recombinant Human SDHA protein (Tagged) (ab226268)

(Tris-Glycine gel) Discontinuous SDS-PAGE (reduced) analysis of ab226268 with 5% enrichment gel and 15% separation gel.



Mass Spectrometry - Recombinant Human SDHA protein (Tagged) (ab226268)

Based on the SEQUEST from database of E.coli host and target protein, the LC-MS/MS analysis result of ab226268 could indicate that this peptide derived from E.coli-expressed Homo sapiens (Human) SDHA.



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