# abcam

## Product datasheet

# Recombinant Human SDHA protein (His tag) ab226453

### 1 图像

#### 描述

产品名称 重组人SDHA蛋白(His tag)

**纯度** > 90 % SDS-PAGE.

表达系统 Escherichia coli

Accession P31040

**蛋白长度** Protein fragment

无动物成分 No

性质 Recombinant

种属 Human

序列 SAKVSDSISAQYPVVDHEFDAVVVGAGGAGLRAAFGLSEAGF

NTACVTKL

FPTRSHTVAAQGGINAALGNMEEDNWRWHFYDTVKGSDWLGD

QDAIHYMT

EQAPAAVVELENYGMPFSRTEDGKIYQRAFGGQSLKFGKGGQ

AHRCCCVA

DRTGHSLLHTLYGRSLRYDTSYFVEYFALDLLMENGECRGVI

**ALCIEDGS** 

IHRIRAKNTVVATGGYGRTYFSCTSAHTSTGDGTAMITRAGL

**PCQDLEFV** 

QFHPTGIYGAGCLITEGCRGEGGILINSQGERFMERYAPVAK

DLASRDVV

SRSMTLEIREGRGCGPEKDHVYLQLHHLPPEQLATRLPGISE

**TAMIFAGV** 

DVTKEPIPVLPTVHYNMGGIPTNYKGQVLRHVNGQDQIVPGL

YACGEAAC

ASVHGANRLGANSLLDLVVFGRACALSIEESCRPGDKVPPIK

**PNAGEESV** 

MNLDKLRFADGSIRTSELRLSMQKSMQNHAAVFRVGSVLQEG

CGKISKLY

GDLKHLKTFDRGMVWNTDLVETLELQNLMLCALQTIYGAEAR

**KESRGAHA** 

REDYKVRIDEYDYSKPIQGQQKKPFEEHWRKHTLSYVDVGTG

KVTLEYRP VIDKTLNEADCATVPPAIRSY

**氨基酸** 44 to 664

标签 His tag N-Terminus

额外的序列信息 N-terminal 6xHis-SUMO tag.

#### 技术指标

Our Abpromise guarantee covers the use of ab226453 in the following tested applications.

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

应用 SDS-PAGE

形式 Liquid

#### 制备和贮存

稳定性和存储 Shipped at 4°C. Upon delivery aliquot. Store at -20°C or -80°C. Avoid freeze / thaw cycle.

pH: 7.2

Constituents: Tris buffer, 50% Glycerol (glycerin, glycerine)

#### 常规信息

功能 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).

通路 Carbohydrate metabolism; tricarboxylic acid cycle; fumarate from succinate (eukaryal route): step

1/1.

疾病相关 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

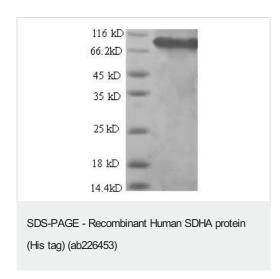
characterized by bilaterally symmetrical necrotic lesions in subcortical brain regions.

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.

序列相似性 Belongs to the FAD-dependent oxidoreductase 2 family. FRD/SDH subfamily.

细**胞定位** Mitochondrion inner membrane.

#### 图片



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

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

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