# abcam

## Product datasheet

## Recombinant Human SGSH/HSS protein ab116779

## 画像数1

#### 製品の詳細

製品名 Recombinant Human SGSH/HSS protein

発現系Wheat germアクセッション番号P51688

タンパク質長 Full length protein

Animal free No

**由来** Recombinant

生物種 Human

配列 MSCPVPACCALLLVLGLCRARPRNALLLLADDGGFESGAYNN

SAIATPHL

DALARRSLLFRNAFTSVSSCSPSRASLLTGLPQHQNGMYGLH

**QDVHHFNS** 

 ${\tt FDKVRSLPLLLSQAGVRTGIIGKKHVGPETVYPFDFAYTEEN}$ 

**GSVLQVGR** 

NITRIKLLVRKFLQTQDDRPFFLYVAFHDPHRCGHSQPQYGT

**FCEKFGNG** 

ESGMGRIPDWTPQAYDPLDVLVPYFVPNTPAARADLAAQYTT

VGRMDQGV

GLVLQELRDAGVLNDTLVIFTSDNGIPFPSGRTNLYWPGTAE

**PLLVSSPE** 

HPKRWGQVSEAYVSLLDLTPTILDWFSIPYPSYAIFGSKTIH

LTGRSLLP

ALEAEPLWATVFGSQSHHEVTMSYPMRSVQHRHFRLVHNLNF

**KMPFPIDQ** 

DFYVSPTFQDLLNRTTAGQPTGWYKDLRHYYYRARWELYDRS

RDPHETQN

LATDPRFAQLLEMLRDQLAKWQWETHDPWVCAPDGVLEEKLS

PQCQPLHN EL

予測される分子量 81 kDa including tags

**領域** 1 to 502

特性

Our <u>Abpromise guarantee</u> covers the use of ab116779 in the following tested applications.

1

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

アプリケーション Western blot

SDS-PAGE

**ELISA** 

製品の状態 Liquid

備考 This product was previously labelled as SGSH.

#### 前処理および保存

保存方法および安定性 Shipped on dry ice. Upon delivery aliquot and store at -80°C. Avoid freeze / thaw cycles.

pH: 8.00

Constituents: 0.3% Glutathione, 0.79% Tris HCI

#### 関連情報

## **関連疾患** Defects in SGSH are the cause of mucopolysaccharidosis type 3A (MPS3A) [MIM:252900]; also

known as Sanfilippo syndrome A. MPS3A is a severe form of mucopolysaccharidosis type 3, an autosomal recessive lysosomal storage disease due to impaired degradation of heparan sulfate. MPS3 is characterized by severe central nervous system degeneration, but only mild somatic disease. Onset of clinical features usually occurs between 2 and 6 years; severe neurologic degeneration occurs in most patients between 6 and 10 years of age, and death occurs typically during the second or third decade of life. MPS3A is characterized by earlier onset, rapid

progression of symptoms and shorter survival.

**配列類似性** Belongs to the sulfatase family.

翻訳後修飾 The conversion to 3-oxoalanine (also known as C-formylglycine, FGly), of a serine or cysteine

residue in prokaryotes and of a cysteine residue in eukaryotes, is critical for catalytic activity.

細胞内局在 Lysosome.

#### 画像



SDS-PAGE - Recombinant Human SGSH/HSS protein (ab116779)

12.5% SDS-PAGE showing ab116779 at approximately 81.29kDa. Stained with Coomassie Blue.

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

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- Extensive multi-media technical resources to help you
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