

Product datasheet

Anti-SGSH antibody ab72617

1 图像

概述

产品名称	Anti-SGSH抗体
描述	小鼠多克隆抗体to SGSH
宿主	Mouse
经测试应用	适用于: WB
种属反应性	与反应: Human
免疫原	SGSH (NP_000190.1, 1 a.a. ~ 502 a.a) full-length human protein.
阳性对照	SGSH transfected 293T cell line

性能

形式	Liquid
存放说明	Shipped at 4°C. Upon delivery aliquot and store at -20°C or -80°C. Avoid repeated freeze / thaw cycles.
存储溶液	Preservative: None Constituents: 1X PBS, pH 7.2
纯度	Protein A purified
克隆	多克隆
同种型	IgG

应用

Our [Abpromise guarantee](#) covers the use of **ab72617** in the following tested applications.

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

应用	Ab评论	说明
WB		1/500 - 1/1000. Predicted molecular weight: 57 kDa.

靶标

疾病相关

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.

序列相似性

Belongs to the sulfatase family.

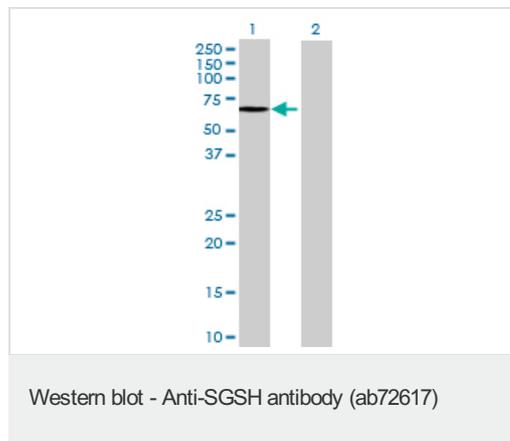
翻译后修饰

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.

细胞定位

Lysosome.

图片



All lanes : Anti-SGSH antibody (ab72617) at 1/500 dilution

Lane 1 : SGSH transfected 293T cell line

Lane 2 : Non transfected 293T cell line

Lysates/proteins at 25 µg per lane.

Secondary

All lanes : Goat Anti-Mouse IgG (H&L)-HRP Conjugate at 1/2500 dilution

Developed using the ECL technique.

Predicted band size: 57 kDa

Observed band size: 67 kDa

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