abcam

Product datasheet

Anti-ARSB antibody ab93123

2 图像

概述

产品名称	Anti-ARSB抗体
描述	兔多克隆抗体to ARSB
宿主	Rabbit
经测试应用	**适用于:** WB, ELISA
种属反应性	**与反应:** Human
免疫原	KLH conjugated synthetic peptide selected from the center region of Human ARSB (NP_000037).
阳性对照	HL-60 cell line lysates; 293 cell lysates transiently transfected with the ARSB gene

性能

形式	Liquid
存放说明	Shipped at 4°C. Store at 4°C (up to 6 months). Store at -20°C long term.
存储溶液	Preservative: 0.09% Sodium azide Constituent: PBS
纯度	Immunogen affinity purified
纯化说明	This antibody is purified through a protein A column, followed by peptide affinity purification.
克隆	多克隆
同种型	IgG

应用

The Abpromise guarantee **Abpromise™承诺保证使用ab93123于以下的经测试应用**

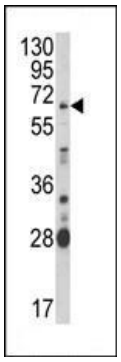
"应用说明"部分 下显示的仅为推荐的起始稀释度;实际最佳的稀释度/浓度应由使用者检定。

应用	Ab评论	说明
WB		1/100 - 1/500. Predicted molecular weight: 60 kDa.
ELISA		1/1000.

靶标

疾病相关	Defects in ARSB are the cause of mucopolysaccharidosis type 6 (MPS6) [MIM:253200]; also known as Maroteaux-Lamy syndrome. MPS6 is an autosomal recessive lysosomal storage disease characterized by intracellular accumulation of dermatan sulfate. Clinical features can include abnormal growth, short stature, stiff joints, skeletal malformations, corneal clouding, hepatosplenomegaly, and cardiac abnormalities. A wide variation in clinical severity is observed. Arylsulfatase B activity is defective in multiple sulfatase deficiency (MSD) [MIM:272200]. MSD is a disorder characterized by decreased activity of all known sulfatases. MSD is due to defects in SUMF1 resulting in the lack of post-translational modification of a highly conserved cysteine into 3-oxoalanine. It combines features of individual sulfatase deficiencies such as metachromatic leukodystrophy, mucopolysaccharidosis, chondrodysplasia punctata, hydrocephalus, ichthyosis, neurologic deterioration and developmental delay.
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. This post-translational modification is severely defective in multiple sulfatase deficiency (MSD).
细胞定位	Lysosome.

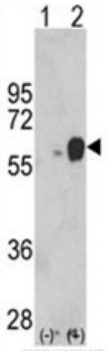
图片



Western blot - Anti-ARSB antibody (ab93123)

Anti-ARSB antibody (ab93123) at 1/100 dilution + HL-60 cell line lysates at 35 µg

Predicted band size: 60 kDa



Western blot - Anti-ARSB antibody (ab93123)

All lanes : Anti-ARSB antibody (ab93123) at 1/100 dilution

Lane 1 : nontransfected 293 cell lysates
Lane 2 : 293 cell lysates transiently transfected with the ARSB gene

Lysates/proteins at 2 µg per lane.

Predicted band size: 60 kDa

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

Our Abpromise to you: Quality guaranteed and expert technical support

- Replacement or refund for products not performing as stated on the datasheet
- Valid for 12 months from date of delivery
- Response to your inquiry within 24 hours
- We provide support in Chinese, English, French, German, Japanese and Spanish
- Extensive multi-media technical resources to help you
- We investigate all quality concerns to ensure our products perform to the highest standards

If the product does not perform as described on this datasheet, we will offer a refund or replacement. For full details of the Abpromise, please visit **https://www.abcam.cn/abpromise** or contact our technical team.

Terms and conditions

- Guarantee only valid for products bought direct from Abcam or one of our authorized distributors