abcam

Product datasheet

Anti-ARSB antibody ab85727

1 References 2 图像

概述

产品名称	Anti-ARSB抗体
描述	**山羊多克隆抗体**to ARSB
宿主	Goat
经测试应用	**适用于:** WB, IHC-P
种属反应性	**与反应:** Human **预测可用于:** Cow, Cat
免疫原	Synthetic peptide corresponding to Human ARSB (internal sequence) (Cysteine residue). Run BLAST with ExPASy Run BLAST with NCBI
阳性对照	Skeletal muscle tissue; human heart lysate.
常规说明	The Life Science industry has been in the grips of a reproducibility crisis for a number of years. Abcam is leading the way in addressing this with our range of recombinant monoclonal antibodies and knockout edited cell lines for gold-standard validation. Please check that this product meets your needs before purchasing. If you have any questions, special requirements or concerns, please send us an inquiry and/or contact our Support team ahead of purchase. Recommended alternatives for this product can be found below, along with publications, customer reviews and Q&As

性能

形式	Liquid
存放说明	Shipped at 4°C. Upon delivery aliquot and store at -20°C. Avoid freeze / thaw cycles.
存储溶液	pH: 7.30 Preservative: 0.02% Sodium azide Constituents: Tris buffered saline, 0.5% BSA
纯度	Immunogen affinity purified
克隆	**多克隆**
同种型	IgG

应用

The Abpromise guarantee **Abpromise™承诺保证使用ab85727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		Use a concentration of 0.03 - 0.1 µg/ml. Predicted molecular weight: 60 kDa.
IHC-P		Use a concentration of 5 µg/ml.

靶标

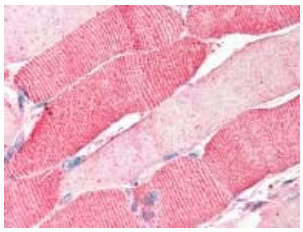
疾病相关 Defects in ARSB are the cause of mucopolysaccharidosis type 6 (MPS6) [MIM:253200]; also known as Maroteaux-Lamy syndrome. MPS6 is an autosomal recessive lysosomal storage disease characterized by intracellular accumulation of dermatan sulfate. Clinical features can include abnormal growth, short 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).

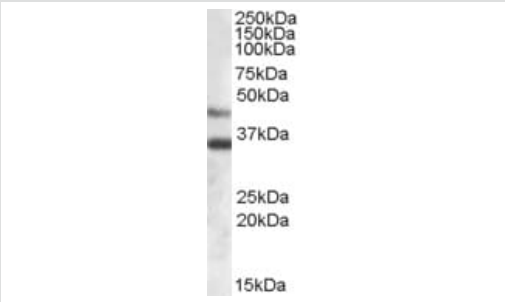
细胞定位 Lysosome.

图片



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-ARSB antibody (ab85727)

ab85727, at 5 µg/ml, staining ARSB in skeletal muscle tissue by Immunohistochemistry.



Western blot - Anti-ARSB antibody (ab85727)

Anti-ARSB antibody (ab85727) at 0.03 µg/ml + human heart lysate at 35 µg

Predicted band size: 60 kDa
Observed band size: 35,40 kDa

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