abcam

Product datasheet

Anti-ARSB antibody ab85727

1 References 画像数 2

製品の概要

製品名 Anti-ARSB antibody

製品の詳細 Goat polyclonal to ARSB

由来種 Goat

アプリケーション 適用あり: WB, IHC-P

種交差性 交差種: Human

交差が予測される動物種: Cow, Cat 🕰

免疫原 Synthetic peptide corresponding to Human ARSB (internal sequence) (Cysteine residue).

ポジティブ・コントロール Skeletal muscle tissue; human heart lysate.

特記事項

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

contact our outport team aread of purchase. Recommended alternatives for this product

found below, along with publications, customer reviews and Q&As

製品の特性

製品の状態 Liquid

保存方法 Shipped at 4°C. Upon delivery aliquot and store at -20°C. Avoid freeze / thaw cycles.

バッファー pH: 7.30

Preservative: 0.02% Sodium azide

Constituents: Tris buffered saline, 0.5% BSA

精製度 Immunogen affinity purified

ポリ/モノ ポリクローナル

アイソタイプ lqG

アプリケーション

The Abpromise guarantee <u>Abpromise保証は、</u>次のテスト済みアプリケーションにおけるab85727の使用に適用されます アプリケーションノートには、推奨の開始希釈率がありますが、適切な希釈率につきましてはご検討ください。

アプリケーション	Abreviews	特記事項
WB		Use a concentration of 0.03 - 0.1 $\mu g/ml$. Predicted molecular weight: 60 kDa.
IHC-P		Use a concentration of 5 µg/ml.

ターゲット情報

関連疾患

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.

配列類似性

翻訳後修飾

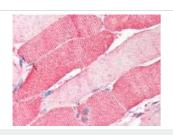
Belongs to the sulfatase family.

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).

細胞内局在

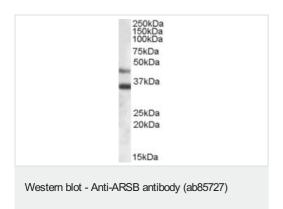
Lysosome.

画像



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

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



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

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

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

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