abcam

Product datasheet

Anti-ARSB antibody - C-terminal ab181410

画像数 1

製品の概要

製品名	Anti-ARSB antibody - C-terminal
製品の詳細	Rabbit polyclonal to ARSB - C-terminal
由来種	Rabbit
特異性	ab181410 only recognizes the longest of the two known isoforms.
アプリケーション	**適用あり:** WB
種交差性	**交差種:** Mouse
免疫原	Synthetic peptide corresponding to Human ARSB (C terminal). Peptide corresponds to 16 amino acids. (NP_000037) Database link: **P15848**
ポジティブ・コントロール	WB: Mouse lung tissue 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 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. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C long term. Avoid freeze / thaw cycle.
バッファー	pH: 7.2 Preservative: 0.02% Sodium azide Constituent: 99% PBS
精製度	Immunogen affinity purified
ポリ/モノ	ポリクローナル
アイソタイプ	IgG

アプリケーション

The Abpromise guarantee **Abpromise保証は、** 次のテスト済みアプリケーションにおけるab181410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 1 - 2 µg/ml. Detects a band of approximately 57 kDa (predicted molecular weight: 60 kDa).

ターゲット情報

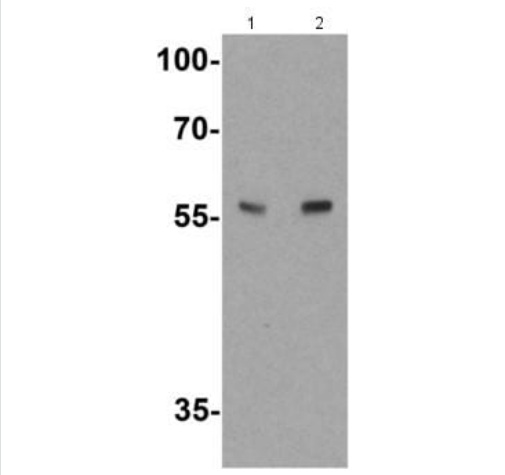
関連疾患 Defects in ARSB are the cause of mucopolysaccharidosis type 6 (MPS6) [MIM:253200]; also known as Maroteaux-Lamy syndrome. MPS6 is an autosomal recessive lysosomal storage disease characterized by intracellular accumulation of dermatan sulfate. Clinical features can include abnormal growth, short 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 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. This post-translational modification is severely defective in multiple sulfatase deficiency (MSD).

細胞内局在 Lysosome.

画像



Western blot - Anti-ARSB antibody - C-terminal (ab181410)

Lane 1 : Anti-ARSB antibody - C-terminal (ab181410) at 1 µg/ml
Lane 2 : Anti-ARSB antibody - C-terminal (ab181410) at 2 µg/ml

All lanes : mouse lung tissue lysate

Lysates/proteins at 15 µg per lane.

Developed using the ECL technique.

Predicted band size: 60 kDa

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

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- We investigate all quality concerns to ensure our products perform to the highest standards

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