abcam

Product datasheet

Anti-ARSB antibody ab115894

1 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
种属反应性	**与反应:** Mouse **预测可用于:** Rat, Horse, Guinea pig, Cow, Cat, Dog, Human, Pig
免疫原	Synthetic peptide corresponding to a region within internal sequence amino acids 300-349 (TDNGGQTRSG GNNWPLRGRK GTLWEGGIRG TGFVASPLLK QKGVKSRELM) of Mouse ARSB (NP_033842). Run BLAST with ExPASy Run BLAST with NCBI
阳性对照	Mouse kidney lysate.

性能

形式	Liquid
存放说明	Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C long term.
存储溶液	Preservative: 0.09% Sodium azide Constituents: 2% Sucrose, PBS
纯度	Immunogen affinity purified
克隆	**多克隆**
同种型	IgG

应用

Our Abpromise guarantee covers the use of **ab115894** 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		Use a concentration of 1 µg/ml. Predicted molecular weight: 60 kDa. Good results were obtained when blocked with 5% non-fat dry milk in 0.05% PBS-T.

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.

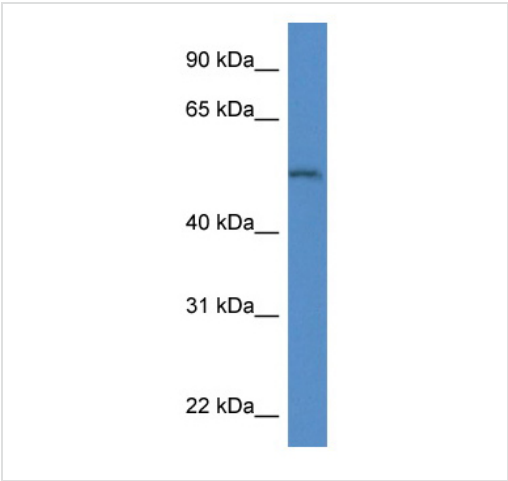
翻译后修饰

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 (ab115894)

Anti-ARSB antibody (ab115894) at 1 µg/ml +
Mouse kidney lysate at 10 µg

Predicted band size: 60 kDa

Gel concentration: 12%

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