abcam

Product datasheet

Anti-ARSA/ASA antibody ab67089

2 图像

概述

产品名称	Anti-ARSA/ASA抗体
描述	**小鼠多克隆抗体**to ARSA/ASA
宿主	Mouse
经测试应用	**适用于:** WB, IHC-P
种属反应性	**与反应:** Human
免疫原	Recombinant full length protein within Human ARSA/ASA aa 1-550. The exact immunogen sequence used to generate this antibody is proprietary information. If additional detail on the immunogen is needed to determine the suitability of the antibody for your needs, please **contact** our Scientific Support team to discuss your requirements.
阳性对照	Lysate from 293T cells transfected with ARSA/ASA.
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 or -80°C. Avoid repeated freeze / thaw cycles.
存储溶液	pH: 7.40 Constituent: 100% PBS
纯度	Protein A purified
克隆	**多克隆**
同种型	IgG

应用

The Abpromise guarantee **Abpromise™承诺保证使用ab67089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 at an assay dependent concentration. Detects a band of approximately 70 kDa (predicted molecular weight: 54 kDa).
IHC-P		Use a concentration of 3 µg/ml.

靶标

功能 Hydrolyzes cerebroside sulfate.

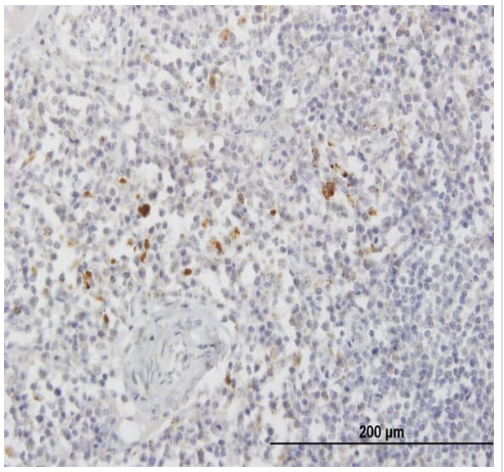
疾病相关 Defects in ARSA are a cause of leukodystrophy metachromatic (MLD) [MIM:250100]. MLD is a disease due to a lysosomal storage defect. It is characterized by intralysosomal storage of cerebroside-3-sulfate in neural and non-neural tissues, with a diffuse loss of myelin in the central nervous system. Progressive demyelination causes a variety of neurological symptoms, including gait disturbances, ataxias, optical atrophy, dementia, seizures, and spastic tetraparesis. Three forms of the disease can be distinguished according to the age at onset: late-infantile, juvenile and adult.
Arylsulfatase A 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).

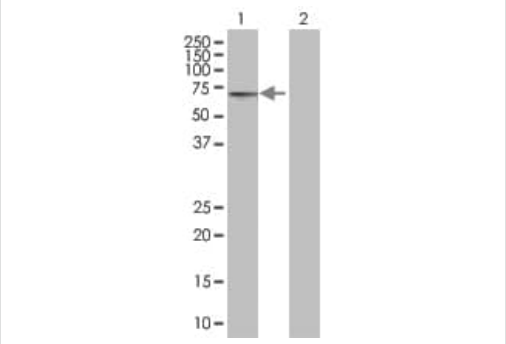
细胞定位 Lysosome.

图片



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-ARSA/ASA antibody (ab67089)

Immunohistochemistry of human lymph node staining ARSA/ASA using ab67089 at 3 µg/ml.



Western blot - Anti-ARSA/ASA antibody (ab67089)

All lanes : Anti-ARSA/ASA antibody (ab67089)

Lane 1 : ARSA transfected 293T cells lysate
Lane 2 : non transfected 293T cells lysate

Secondary
All lanes : Goat Anti-Mouse IgG (H&L)-HRP Conjugated at 1/2500 dilution

Predicted band size: 54 kDa
Observed band size: 70 kDa

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