


### Anti-ARSA/ASA antibody ab154300

画像 1

医薬用外劇物

#### 製品の概要

製品名	Anti-ARSA/ASA antibody
製品の詳細	Rabbit polyclonal to ARSA/ASA
由来種	Rabbit
アプリケーション	適用あり: WB
種交差性	交差種: Human 交差が予測される動物種: Mouse, Rat, Cow, Dog 
免疫原	Recombinant fragment corresponding to Human ARSA/ASA aa 169-405 (internal sequence).
ポジティブ・コントロール	A431; H1299; HeLa and HepG2 cell lysates.
特記事項	<p>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.</p> <p>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&amp;As</p>

#### 製品の特性

製品の状態	Liquid
保存方法	Shipped at 4°C. Upon delivery aliquot. Store at -20°C or -80°C. Avoid freeze / thaw cycle.
バッファー	pH: 7.00 Preservative: 0.01% Thimerosal (merthiolate) Constituents: 1.21% Tris, 0.75% Glycine, 10% Glycerol (glycerin, glycerine)
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ab154300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確認ください。

アプリケーション	Abreviews	特記事項
WB		1/500 - 1/3000. Predicted molecular weight: 54 kDa.

## ターゲット情報

### 機能

Hydrolyzes cerebroside sulfate.

### 関連疾患

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.

### 配列類似性

Belongs to the sulfatase family.

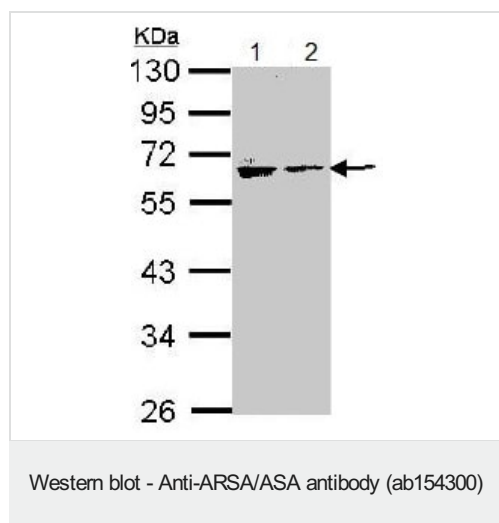
### 翻訳後修飾

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



**All lanes :** Anti-ARSA/ASA antibody (ab154300) at 1/1000 dilution

**Lane 1 :** H1299 whole cell lysate

**Lane 2 :** HeLa whole cell lysate

Lysates/proteins at 30 µg per lane.

**Predicted band size:** 54 kDa

10% SDS PAGE

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

### **Our Abpromise to you: Quality guaranteed and expert technical support**

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- Replacement or refund for products not performing as stated on the datasheet
- Valid for 12 months from date of delivery
- Response to your inquiry within 24 hours
  
- We provide support in Chinese, English, French, German, Japanese and Spanish
- Extensive multi-media technical resources to help you
- We investigate all quality concerns to ensure our products perform to the highest standards

If the product does not perform as described on this datasheet, we will offer a refund or replacement. For full details of the Abpromise, please visit <https://www.abcam.co.jp/abpromise> or contact our technical team.

### **Terms and conditions**

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- Guarantee only valid for products bought direct from Abcam or one of our authorized distributors