abcam

Product datasheet

Anti-ARSA/ASA antibody ab154300

1 Image

Overview

Product name	Anti-ARSA/ASA antibody
Description	Rabbit polyclonal to ARSA/ASA
Host species	Rabbit
Tested applications	**Suitable for:** WB
Species reactivity	**Reacts with:** Human **Predicted to work with:** Mouse, Rat, Cow, Dog
Immunogen	Recombinant fragment corresponding to Human ARSA/ASA aa 169-405 (internal sequence).
Positive control	A431; H1299; HeLa and HepG2 cell lysates.
General notes	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

Properties

Form	Liquid
Storage instructions	Shipped at 4°C. Upon delivery aliquot. Store at -20°C or -80°C. Avoid freeze / thaw cycle.
Storage buffer	pH: 7.00 Preservative: 0.01% Thimerosal (merthiolate) Constituents: 1.21% Tris, 0.75% Glycine, 10% Glycerol (glycerin, glycerine)
Purity	Immunogen affinity purified
Clonality	Polyclonal
Isotype	IgG

Applications

The Abpromise guarantee	Our **Abpromise guarantee** covers the use of ab154300 in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Application	Abreviews	Notes
WB		1/500 - 1/3000. Predicted molecular weight: 54 kDa.

Target

Function

Hydrolyzes cerebroside sulfate.

Involvement in disease

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.

Sequence similarities

Belongs to the sulfatase family.

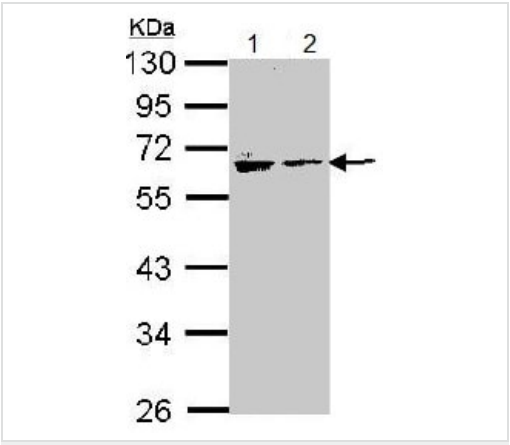
Post-translational modifications

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).

Cellular localization

Lysosome.

Images



Western blot - Anti-ARSA/ASA antibody (ab154300)

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

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