abcam

Product datasheet

Anti-ARSB antibody - C-terminal ab82416

1 Image

Overview

Product name	Anti-ARSB antibody - C-terminal
Description	Rabbit polyclonal to ARSB - C-terminal
Host species	Rabbit
Tested applications	**Suitable for:** WB, ELISA
Species reactivity	**Reacts with:** Human
Immunogen	KLH conjugated synthetic peptide selected from the C-terminal region of human ARSB.
Positive control	HepG2 cell lysate.

Properties

Form	Liquid
Storage instructions	Shipped at 4°C. Upon delivery aliquot and store at -20°C. Avoid freeze / thaw cycles.
Storage buffer	Preservative: 0.09% Sodium Azide Constituents: PBS
Purity	Ammonium Sulphate Precipitation
Purification notes	ab82416 is prepared by Saturated Ammonium Sulfate (SAS) precipitation followed by dialysis against PBS.
Clonality	Polyclonal
Isotype	IgG

Applications

Our Abpromise guarantee covers the use of **ab82416** in the following tested applications.

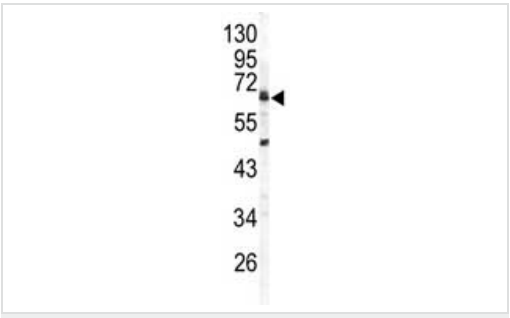
The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Application	Abreviews	Notes
WB		1/50 - 1/100. Predicted molecular weight: 60 kDa.
ELISA		1/1000.

Target

Involvement in disease	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.
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-ARSB antibody - C-terminal (ab82416)

Anti-ARSB antibody - C-terminal (ab82416) at 1/50 dilution + HepG2 lysate at 35 µg

Predicted band size: 60 kDa
Observed band size: 65 kDa
Additional bands at: 50 kDa. We are unsure as to the identity of these extra bands.

Our Abpromise to you: Quality guaranteed and expert technical support

- 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.com/abpromise or contact our technical team.

Terms and conditions

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