

Product datasheet

Anti-ARSB antibody - C-terminal ab82416

1 Image

Overview

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

Properties

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

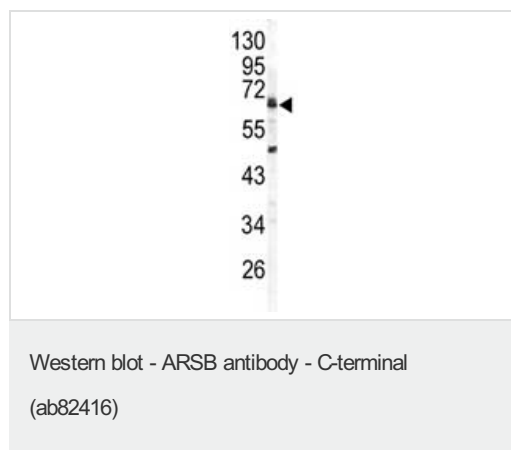
The conversion to 3-oxoalanine (also known as C-formylglycine, FGly), of a serine or cysteine residue in prokaryotes and of a cysteine residue in eukaryotes, is critical for catalytic activity.

This post-translational modification is severely defective in multiple sulfatase deficiency (MSD).

### Cellular localization

Lysosome.

## Images



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.

**Please note:** All products are "FOR RESEARCH USE ONLY AND ARE NOT INTENDED FOR DIAGNOSTIC OR THERAPEUTIC USE"

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