Product datasheet

Anti-Acid sphingomyelinase antibody ab83354

2 Abreviews 10 References 3 Images

Overview

Product name: Anti-Acid sphingomyelinase antibody
Description: Rabbit polyclonal to Acid sphingomyelinase
Host species: Rabbit
Tested applications: Suitable for: ICC/IF, WB, IHC-P
Species reactivity: Reacts with: Human
          Predicted to work with: Mouse, Rat, Sheep, Rabbit, Horse, Guinea pig, Cow, Cat, Dog, Pig

Immunogen: Synthetic peptide corresponding to a region within internal sequence amino acids 396-445 (INSTDPAGQL QWLVEGLQAA EDRGDKVHII GHIPPGHCLK SWSWNYYRM) of Human Acid sphingomyelinase (NP_000534).

Positive control: Fetal heart lysate.

Properties

Form: Liquid
Storage instructions: Shipped at 4°C. Upon delivery aliquot and store at -20°C. Avoid repeated freeze / thaw cycles.
Storage buffer: Preservative: 0.09% Sodium azide
          Constituents: 2% Sucrose, PBS
Purity: Immunogen affinity purified
Clonality: Polyclonal
Isotype: IgG

Applications

Our Abpromise guarantee covers the use of ab83354 in the following tested applications.
The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

<table>
<thead>
<tr>
<th>Application</th>
<th>Abreviews</th>
<th>Notes</th>
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<tbody>
<tr>
<td>ICC/IF</td>
<td></td>
<td>Use a concentration of 1 µg/ml.</td>
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</table>
Function
Converts sphingomyelin to ceramide. Also has phospholipase C activities toward 1,2-diacylglycerolphosphocholine and 1,2-diacylglycerolphosphoglycerol. Isoform 2 and isoform 3 have lost catalytic activity.

Involvement in disease
Defects in SMPD1 are the cause of Niemann-Pick disease type A (NPDA) [MIM:257200]; also known as Niemann-Pick disease classical infantile form. It is an early-onset lysosomal storage disorder caused by failure to hydrolyze sphingomyelin to ceramide. It results in the accumulation of sphingomyelin and other metabolically related lipids in reticuloendothelial and other cell types throughout the body, leading to cell death. Niemann-Pick disease type A is a primarily neurodegenerative disorder characterized by onset within the first year of life, mental retardation, digestive disorders, failure to thrive, major hepatosplenomegaly, and severe neurologic symptoms. The severe neurological disorders and pulmonary infections lead to an early death, often around the age of four. Clinical features are variable. A phenotypic continuum exists between type A (basic neurovisceral) and type B (purely visceral) forms of Niemann-Pick disease, and the intermediate types encompass a cluster of variants combining clinical features of both types A and B.

Defects in SMPD1 are the cause of Niemann-Pick disease type B (NPDB) [MIM:607616]; also known as Niemann-Pick disease visceral form. It is a late-onset lysosomal storage disorder caused by failure to hydrolyze sphingomyelin to ceramide. It results in the accumulation of sphingomyelin and other metabolically related lipids in reticuloendothelial and other cell types throughout the body, leading to cell death. Clinical signs involve only visceral organs. The most constant sign is hepatosplenomegaly which can be associated with pulmonary symptoms. Patients remain free of neurologic manifestations. However, a phenotypic continuum exists between type A (basic neurovisceral) and type B (purely visceral) forms of Niemann-Pick disease, and the intermediate types encompass a cluster of variants combining clinical features of both types A and B. In Niemann-Pick disease type B, onset of the first symptoms occurs in early childhood and patients can survive into adulthood.

Sequence similarities
Belongs to the acid sphingomyelinase family.
Contains 1 saposin B-type domain.

Cellular localization
Lysosome.

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<tr>
<td>WB</td>
<td>⭐⭐⭐⭐⭐</td>
<td>Use a concentration of 1 µg/ml. Predicted molecular weight: 70 kDa. Good results were obtained when blocked with 5% non-fat dry milk in 0.05% PBS-T.</td>
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<tr>
<td>IHC-P</td>
<td></td>
<td>Use a concentration of 1 µg/ml. Perform heat mediated antigen retrieval with citrate buffer pH 6 before commencing with IHC staining protocol.</td>
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</tbody>
</table>

Images
Western blot - Anti-Acid sphingomyelinase antibody (ab83354) at 1 µg/ml (in 5% skim milk / PBS buffer) + Human fetal heart lysate at 10 µg

**Secondary**
HRP conjugated anti-Rabbit IgG at 1/50000 dilution

**Predicted band size:** 70 kDa

**Observed band size:** 65 kDa

*why is the actual band size different from the predicted?*

**Additional bands at:** 58 kDa. We are unsure as to the identity of these extra bands.

**Gel concentration:** 12%

ICC/IF image of ab83354 stained HeLa cells. The cells were 4% formaldehyde fixed (10 min) and then incubated in 1%BSA / 10% normal goat serum / 0.3M glycine in 0.1% PBS-Tween for 1h to permeabilise the cells and block non-specific protein-protein interactions. The cells were then incubated with the antibody (ab83354, 1µg/ml) overnight at +4°C. The secondary antibody (green) was Alexa Fluor® 488 goat anti-rabbit IgG (H+L) used at a 1/1000 dilution for 1h. Alexa Fluor® 594 WGA was used to label plasma membranes (red) at a 1/200 dilution for 1h. DAPI was used to stain the cell nuclei (blue) at a concentration of 1.43µM.
Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Acid sphingomyelinase antibody (ab83354)

IHC image of ab83354 staining in human normal cervix formalin fixed paraffin embedded tissue section, performed on a Leica Bond™ system using the standard protocol F. The section was pre-treated using heat mediated antigen retrieval with sodium citrate buffer (pH6, epitope retrieval solution 1) for 20 mins. The section was then incubated with ab83354, 1µg/ml, for 15 mins at room temperature and detected using an HRP conjugated compact polymer system. DAB was used as the chromogen. The section was then counterstained with haematoxylin and mounted with DPX.

For other IHC staining systems (automated and non-automated) customers should optimize variable parameters such as antigen retrieval conditions, primary antibody concentration and antibody incubation times.

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