abcam

Product datasheet

Anti-Niemann Pick C1 antibody [EPR5209] - BSA and Azide free ab224268

敲除验证 重组 RabMAb

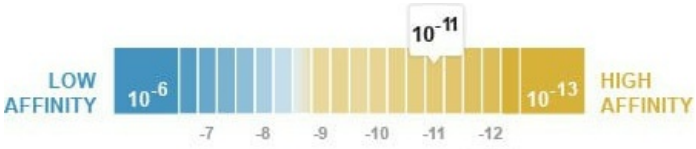
1 References 11 图像

概述

产品名称	Anti-Niemann Pick C1抗体[EPR5209] - BSA and Azide free
描述	兔单克隆抗体[EPR5209] to Niemann Pick C1 - BSA and Azide free
宿主	Rabbit
经测试应用	**适用于:** Flow Cyt (Intra), ICC/IF, WB, IHC-P
种属反应性	**与反应:** Mouse, Rat, Human
免疫原	Synthetic peptide. This information is proprietary to Abcam and/or its suppliers.
阳性对照	3T3L1 cell lysate, L6 cell lysate, HepG2 cell lysate, THP1 cell lysate, 293T cell lysate, PC3 cell lysate, Rat liver lysate, Rat brain lysate, Human kidney tissue.
常规说明	ab224268 is the carrier-free version of **ab134113**. Our **carrier-free** antibodies are typically supplied in a PBS-only formulation, purified and free of BSA, sodium azide and glycerol. The carrier-free buffer and high concentration allow for increased conjugation efficiency. This conjugation-ready format is designed for use with fluorochromes, metal isotopes, oligonucleotides, and enzymes, which makes them ideal for antibody labelling, functional and cell-based assays, flow-based assays (e.g. mass cytometry) and Multiplex Imaging applications. Use our **conjugation kits** for antibody conjugates that are ready-to-use in as little as 20 minutes with <1 minute hands-on-time and 100% antibody recovery: available for fluorescent dyes, HRP, biotin and gold. This product is compatible with the Maxpar® Antibody Labeling Kit from Fluidigm, without the need for antibody preparation. Maxpar® is a trademark of Fluidigm Canada Inc. This product is a recombinant monoclonal antibody, which offers several advantages including: - High batch-to-batch consistency and reproducibility - Improved sensitivity and specificity - Long-term security of supply - Animal-free production For more information **see here**. Our RabMAb® technology is a patented hybridoma-based technology for making rabbit monoclonal antibodies. For details on our patents, please refer to **RabMAb® patents**.

性能

形式	Liquid
存放说明	Shipped at 4°C. Store at +4°C. Do Not Freeze.
解离常数（K_D）	$K_D = 4.90 \times 10^{-11}$ M



Learn more about K_D

存储溶液	pH: 7.2 Constituent: PBS
无载体	是
纯度	Protein A purified
克隆	单克隆
克隆编号	EPR5209
同种型	IgG

应用

The Abpromise guarantee Abpromise™承诺保证使用ab224268于以下的经测试应用

"应用说明"部分 下显示的仅为推荐的起始稀释度;实际最佳的稀释度/浓度应由使用者检定。

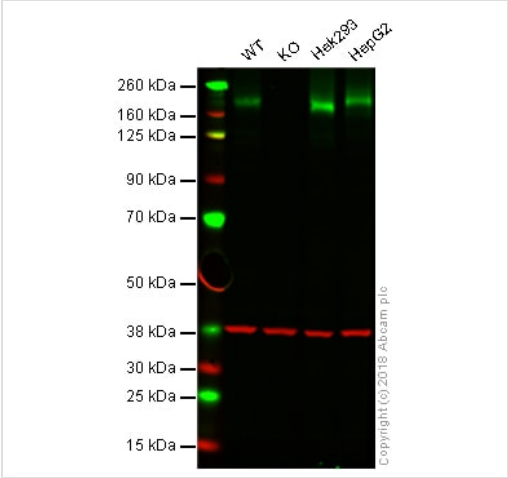
应用	Ab评论	说明
Flow Cyt (Intra)		Use at an assay dependent concentration. **ab199376** - Rabbit monoclonal IgG, is suitable for use as an isotype control with this antibody.
ICC/IF		Use at an assay dependent concentration.
WB		Use at an assay dependent concentration. Detects a band of approximately 180 kDa (predicted molecular weight: 142 kDa).
IHC-P		Use at an assay dependent concentration. Perform heat mediated antigen retrieval with citrate buffer pH 6 before commencing with IHC staining protocol.

靶标

功能	Involved in the intracellular trafficking of cholesterol. May play a role in vesicular trafficking in glia, a process that may be crucial for maintaining the structural and functional integrity of nerve terminals.
疾病相关	Defects in NPC1 are the cause of Niemann-Pick disease type C1 (NPDC1) [MIM:257220]. A lysosomal storage disorder that affects the viscera and the central nervous system. It is due to

	defective intracellular processing and transport of low-density lipoprotein derived cholesterol. It causes accumulation of cholesterol in lysosomes, with delayed induction of cholesterol homeostatic reactions. Niemann-Pick disease type C1 has a highly variable clinical phenotype. Clinical features include variable hepatosplenomegaly and severe progressive neurological dysfunction such as ataxia, dystonia and dementia. The age of onset can vary from infancy to late adulthood. An allelic variant of Niemann-Pick disease type C1 is found in people with Nova Scotia ancestry. Patients with the Nova Scotian clinical variant are less severely affected.
序列相似性	Belongs to the patched family. Contains 1 SSD (sterol-sensing) domain.
结构域	A cysteine-rich N-terminal domain and a C-terminal domain containing a di-leucine motif necessary for lysosomal targeting are critical for mobilization of cholesterol from lysosomes.
翻译后修饰	Glycosylated.
细胞定位	Late endosome membrane. Lysosome membrane.

图片



Western blot - Anti-Niemann Pick C1 antibody [EPR5209] - BSA and Azide free (ab224268)

All lanes : Anti-Niemann Pick C1 antibody [EPR5209] (**ab134113**) at 1/1000 dilution

Lane 1 : Wild-type HAP1 whole cell lysate
Lane 2 : NPC1 (Niemann Pick C1) knockout HAP1 whole cell lysate
Lane 3 : HEK293 whole cell lysate
Lane 4 : HepG2 whole cell lysate

Lysates/proteins at 20 µg per lane.

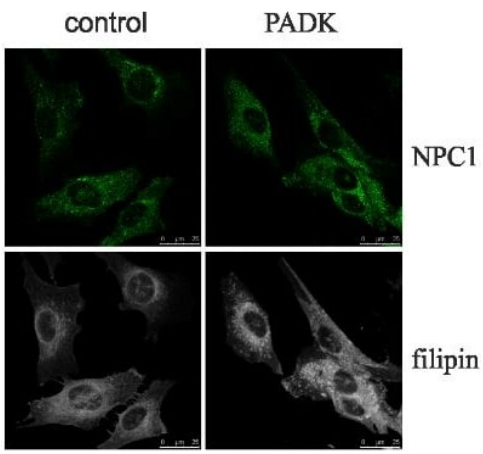
Predicted band size: 142 kDa

Lanes 1 - 4: Merged signal (red and green). Green - **ab134113** observed at 180 kDa. Red - loading control, **ab9484**, observed at 37 kDa.

ab134113 was shown to specifically react with Niemann Pick C1 in wild-type HAP1 cells as signal was lost in NPC1 (Niemann Pick C1) knockout cells. Wild-type and NPC1 (Niemann Pick C1) knockout samples were subjected to SDS-PAGE. Ab134113 and **ab9484** (Mouse anti-GAPDH loading control) were incubated overnight at 4°C at 1/1000 dilution and 1/20000 dilution respectively. Blots were developed with Goat anti-Rabbit IgG H&L (IRDye® 800CW) preabsorbed **ab216773** and Goat anti-Mouse IgG H&L (IRDye® 680RD) preabsorbed **ab216776** secondary antibodies at 1/20000 dilution for 1 hour at room temperature

before imaging.

This data was developed using the same antibody clone in a different buffer formulation containing PBS, BSA, glycerol, and sodium azide (**ab134113**).



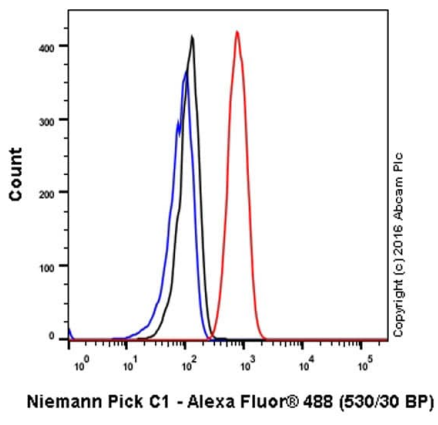
Immunocytochemistry/ Immunofluorescence - Anti-Niemann Pick C1 antibody [EPR5209] - BSA and Azide free (ab224268)

Image from Cermak S et al. PLoS One. 2016;11(11):e0167428. Fig 3.; doi: 10.1371/journal.pone.0167428.

Cathepsin B/L inhibition causes NPC disease-like cholesterol accumulation in SH-SY5Y cells.

Confocal microscopy of SH-SY5Y control and PADK treated cells. Cholesterol (filipin staining, white) and NPC1 (**ab134113**; green).

This data was developed using the same antibody clone in a different buffer formulation containing PBS, BSA, glycerol, and sodium azide (**ab134113**).



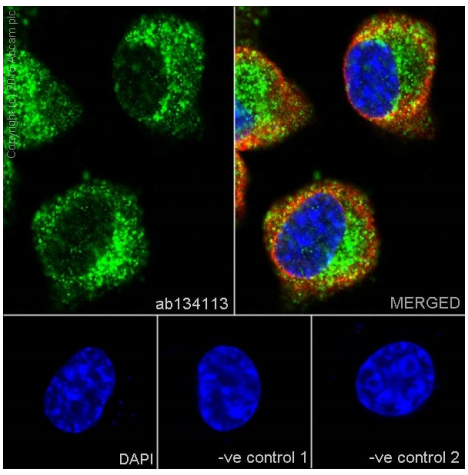
Flow Cytometry (Intracellular) - Anti-Niemann Pick C1 antibody [EPR5209] - BSA and Azide free (ab224268)

ab134113 staining Niemann Pick C1 in Neuro-2a (mouse neuroblastoma cell line) by intracellular flow cytometry. Cells were fixed with 4% paraformaldehyde, permabilised with 90% methanol and the sample was incubated with the primary antibody at a dilution of 1/200. A goat anti rabbit IgG (Alexa Fluor® 488) at a dilution of 1/2000 was used as the secondary antibody.

Isoytype control: Rabbit monoclonal IgG (Black)

Unlabeled control: Cell without incubation with primary antibody and secondary antibody (Blue)

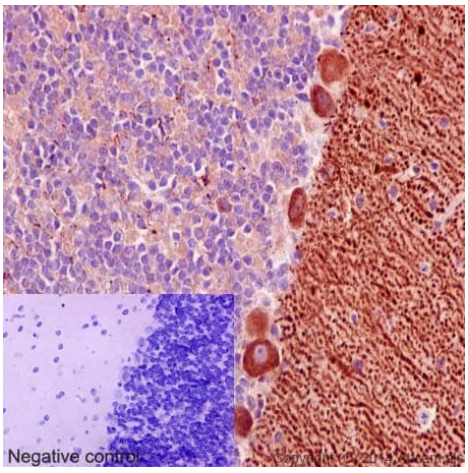
This data was developed using the same antibody clone in a different buffer formulation containing PBS, BSA, glycerol, and sodium azide (**ab134113**).



Immunocytochemistry/ Immunofluorescence - Anti-Niemann Pick C1 antibody [EPR5209] - BSA and Azide free (ab224268)

Immunofluorescence staining of Neuro-2a (mouse neuroblastoma cell line) cells with purified **ab134113** at a working dilution of 1 in 70, counter-stained with DAPI. The secondary antibody was Alexa Fluor® 488 goat anti rabbit (**ab150077**), used at a dilution of 1 in 500. The cells were fixed in 4% PFA and permeabilized using 0.1% Triton X 100. The negative control is shown in bottom right hand panel - for the negative control, purified **ab134113** was used at a dilution of 1/200 followed by an Alexa Fluor® 594 goat anti-mouse antibody (**ab150120**) at a dilution of 1/500.

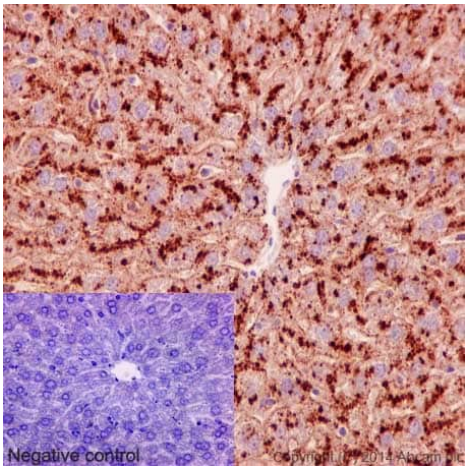
This data was developed using the same antibody clone in a different buffer formulation containing PBS, BSA, glycerol, and sodium azide (**ab134113**).



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Niemann Pick C1 antibody [EPR5209] - BSA and Azide free (ab224268)

Immunohistochemical staining of paraffin embedded rat cerebellum with purified **ab134113** at a working dilution of 1 in 50. The secondary antibody used is a HRP polymer for rabbit IgG. The sample is counter-stained with hematoxylin. Antigen retrieval was perfomed using Tris-EDTA buffer, pH 9.0. PBS was used instead of the primary antibody as the negative control, and is shown in the inset.

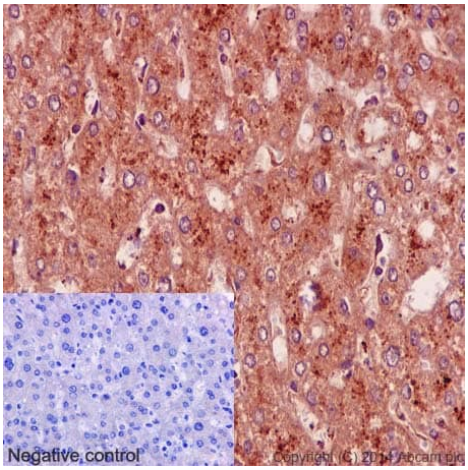
This data was developed using the same antibody clone in a different buffer formulation containing PBS, BSA, glycerol, and sodium azide (**ab134113**).



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Niemann Pick C1 antibody [EPR5209] - BSA and Azide free (ab224268)

Immunohistochemical staining of paraffin embedded mouse liver with purified **ab134113** at a working dilution of 1 in 50. The secondary antibody used is a HRP polymer for rabbit IgG. The sample is counter-stained with hematoxylin. Antigen retrieval was perfomed using Tris-EDTA buffer, pH 9.0. PBS was used instead of the primary antibody as the negative control, and is shown in the inset.

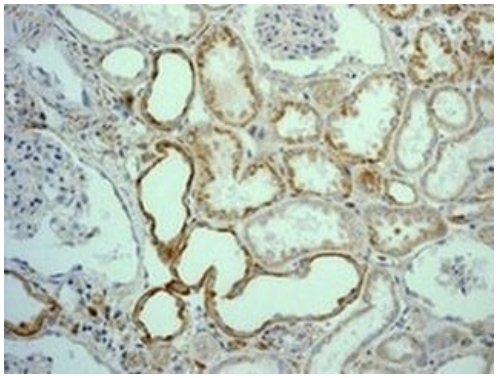
This data was developed using the same antibody clone in a different buffer formulation containing PBS, BSA, glycerol, and sodium azide (**ab134113**).



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Niemann Pick C1 antibody [EPR5209] - BSA and Azide free (ab224268)

Immunohistochemical staining of paraffin embedded human liver with purified **ab134113** at a working dilution of 1 in 50. The secondary antibody used is a HRP polymer for rabbit IgG. The sample is counter-stained with hematoxylin. Antigen retrieval was perfomed using Tris-EDTA buffer, pH 9.0. PBS was used instead of the primary antibody as the negative control, and is shown in the inset.

This data was developed using the same antibody clone in a different buffer formulation containing PBS, BSA, glycerol, and sodium azide (**ab134113**).

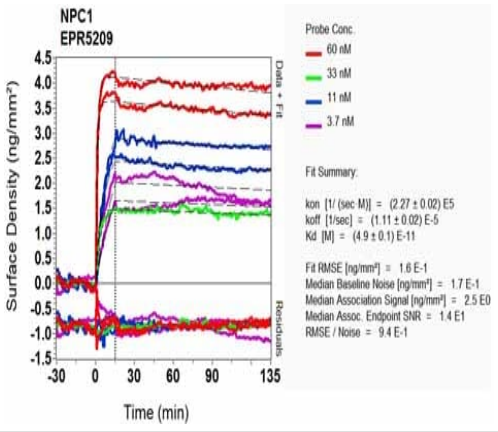


Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Niemann Pick C1 antibody [EPR5209] - BSA and Azide free (ab224268)

Immunohistochemical analysis of paraffin embedded human kidney tissue labelling Niemann Pick C1 with unpurified **ab134113** at 1/50.

This data was developed using the same antibody clone in a different buffer formulation containing PBS, BSA, glycerol, and sodium azide (**ab134113**).

Perform heat mediated antigen retrieval with citrate buffer pH 6 before commencing with IHC staining protocol.



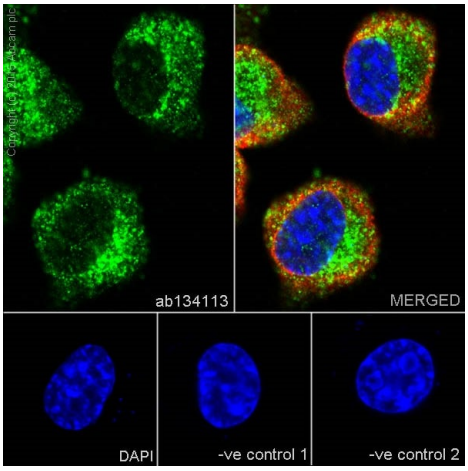
OI-RD Scanning - Anti-Niemann Pick C1 antibody [EPR5209] - BSA and Azide free (ab224268)

Equilibrium disassociation constant (K_D)

Learn more about K_D

Click here to learn more about K_D

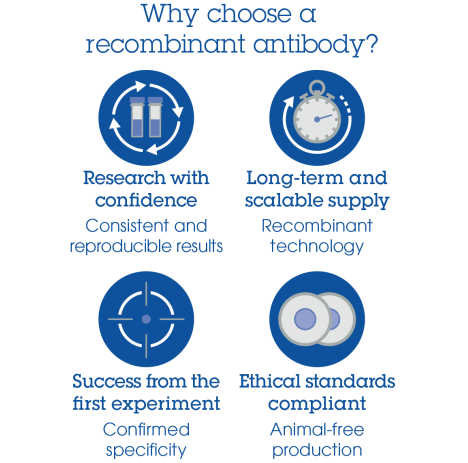
This data was developed using the same antibody clone in a different buffer formulation containing PBS, BSA, glycerol, and sodium azide (**ab134113**).



Immunocytochemistry/ Immunofluorescence - Anti-Niemann Pick C1 antibody [EPR5209] - BSA and Azide free (ab224268)

This ICC data was generated using the same anti-Niemann Pick C1 antibody clone [EPR5209] in a different buffer formulation (cat# **ab134133**).

Immunofluorescence staining of neuro-2a cells with purified **ab134113** at a working dilution of 1 in 70, counter-stained with DAPI. The secondary antibody was Alexa Fluor® 488 goat anti rabbit (**ab150077**), used at a dilution of 1 in 500. The cells were fixed in 4% PFA and permeabilized using 0.1% Triton X 100. The negative control is shown in bottom right hand panel - for the negative control, purified **ab134113** was used at a dilution of 1/200 followed by an Alexa Fluor® 594 goat anti-mouse antibody (**ab150120**) at a dilution of 1/500.



Anti-Niemann Pick C1 antibody [EPR5209] - BSA and Azide free (ab224268)

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