

## Product datasheet

# Anti-Niemann Pick C1 antibody [EPR5209] ab134113

**KO VALIDATED** Recombinant RabMAb

[11 References](#) [12 Images](#)

### Overview

<b>Product name</b>	Anti-Niemann Pick C1 antibody [EPR5209]
<b>Description</b>	Rabbit monoclonal [EPR5209] to Niemann Pick C1
<b>Host species</b>	Rabbit
<b>Tested applications</b>	<b>Suitable for:</b> Flow Cyt, WB, IHC-P, ICC/IF
<b>Species reactivity</b>	<b>Reacts with:</b> Mouse, Rat, Human
<b>Immunogen</b>	Synthetic peptide within Human Niemann Pick C1 aa 1250-1350 (C terminal). The exact sequence is proprietary. Database link: <a href="#">O15118</a>
<b>Positive control</b>	WB: Wild-type HAP1 whole cell lysate; HEK-293, HepG2, THP-1, PC-3, 3T3L1, L6 and HEK-293T cell lysates; Rat liver and brain lysates. IHC-P: Human kidney and liver tissues; Mouse liver tissue; Rat cerebellum tissue. ICC/IF: Neuro-2a cells; SH-SY5Y control and PADK treated cells. Flow Cyt: Neuro-2a cells.

### General notes

Our RabMAb<sup>®</sup> technology is a patented hybridoma-based technology for making rabbit monoclonal antibodies. For details on our patents, please refer to [RabMAb<sup>®</sup> patents](#).

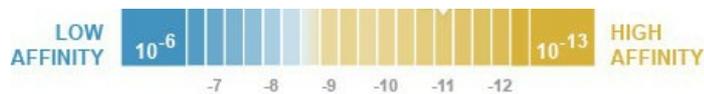
**We are constantly working hard to ensure we provide our customers with best in class antibodies. As a result of this work we are pleased to now offer this antibody in purified format. We are in the process of updating our datasheets. The purified format is designated 'PUR' on our product labels. If you have any questions regarding this update, please contact our Scientific Support team.**

This product is a [recombinant rabbit monoclonal antibody](#).

### Properties

<b>Form</b>	Liquid
<b>Storage instructions</b>	Shipped at 4°C. Store at -20°C. Stable for 12 months at -20°C.
<b>Dissociation constant (K<sub>D</sub>)</b>	K <sub>D</sub> = 4.90 x 10 <sup>-11</sup> M





[Learn more about K<sub>D</sub>](#)

<b>Storage buffer</b>	pH: 7.40 Preservative: 0.01% Sodium azide Constituents: 40% Glycerol, 0.05% BSA, 59% PBS
<b>Purity</b>	Protein A purified
<b>Clonality</b>	Monoclonal
<b>Clone number</b>	EPR5209
<b>Isotype</b>	IgG

## Applications

Our [Abpromise guarantee](#) covers the use of **ab134113** 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
Flow Cyt		1/200. <a href="#">ab172730</a> - Rabbit monoclonal IgG, is suitable for use as an isotype control with this antibody.
WB		1/2000 - 1/10000. Detects a band of approximately 180 kDa (predicted molecular weight: 142 kDa).
IHC-P		1/50 - 1/100. Perform heat mediated antigen retrieval with citrate buffer pH 6 before commencing with IHC staining protocol.
ICC/IF		1/70.

## Target

<b>Function</b>	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.
<b>Involvement in disease</b>	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.
<b>Sequence similarities</b>	Belongs to the patched family. Contains 1 SSD (sterol-sensing) domain.
<b>Domain</b>	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.

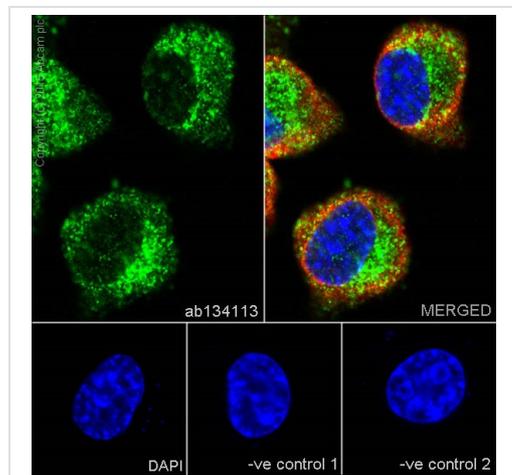
**Post-translational modifications**

Glycosylated.

**Cellular localization**

Late endosome membrane. Lysosome membrane.

**Images**



Immunocytochemistry/ Immunofluorescence - Anti-Niemann Pick C1 antibody [EPR5209] (ab134113)

Immunofluorescent analysis of 4% paraformaldehyde-fixed, 0.1% Triton X-100 permeabilized Neuro-2a (mouse neuroblastoma cell line) cells labeling Niemann Pick C1 with ab134113 at 1/70 followed by Goat anti-rabbit IgG (Alexa Fluor® 488) (ab150077) secondary antibody at 1/500 (green). Confocal image showing cytoplasmic staining on Neuro-2a cell line. The nuclear counterstain is DAPI (blue). Tubulin is detected with ab7291 (anti-Tubulin mouse mAb) at 1/1000 and ab150120 (Alexa Fluor®594 Goat anti-Mouse secondary) at 1/500 (red).

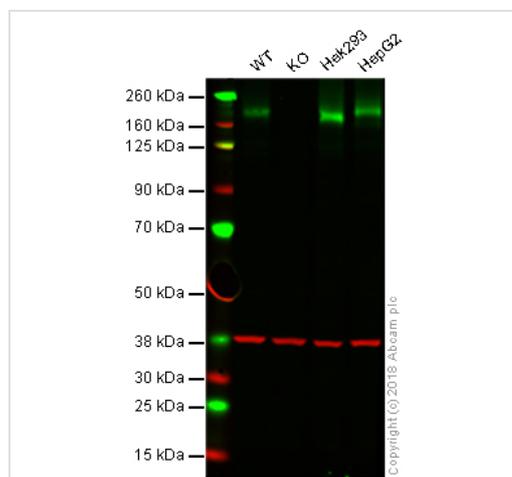
The negative controls are as follows:

-ve control 1 – ab134113 at 1/70 followed

by ab150120 (Alexa Fluor®594 Goat anti-Mouse secondary) at 1/500.

-ve control 2. - ab7291 (anti-Tubulin mouse mAb) at 1/1000

followed by ab150077 (Alexa Fluor®488 Goat Anti-Rabbit IgG H&L) at 1/500.



Western blot - Anti-Niemann Pick C1 antibody [EPR5209] (ab134113)

**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

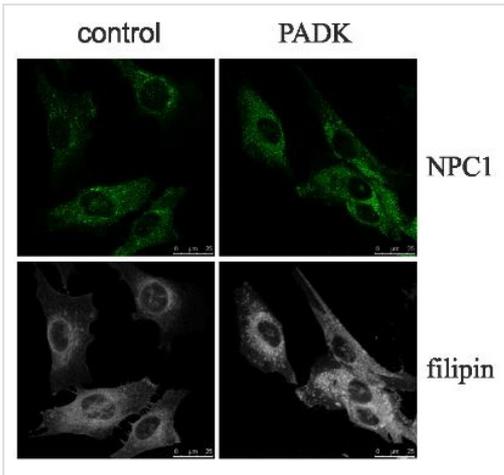
Lysates/proteins at 20 µg per lane.

**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.

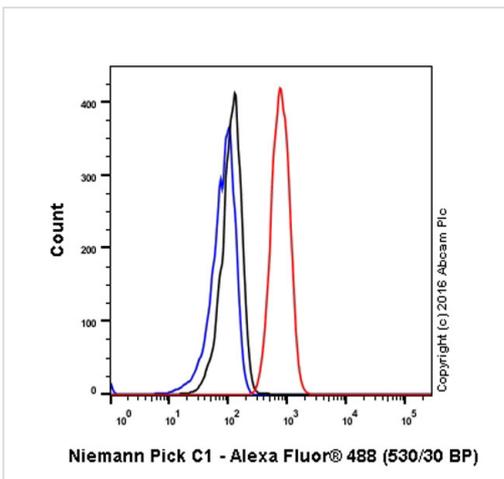


**Cathepsin B/L inhibition causes NPC disease-like cholesterol accumulation in SH-SY5Y cells.**

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

Immunocytochemistry/ Immunofluorescence - Anti-Niemann Pick C1 antibody [EPR5209] (ab134113)

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

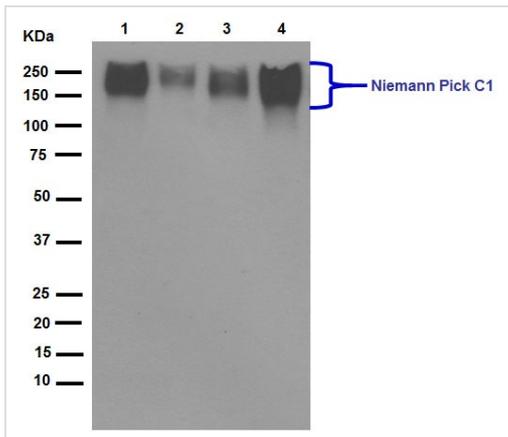


ab134113 staining Niemann Pick C1 in Neuro-2a (mouse neuroblastoma cell line) by flow cytometry. Cells were fixed with 4% paraformaldehyde, permabilised with 90% methanol and the sample was incubated with the primary antibody at a dilution of 1/200. A goat anti rabbit IgG (Alexa Fluor® 488) at a dilution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

Isotype control: Rabbit monoclonal IgG (Black)

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

Flow Cytometry - Anti-Niemann Pick C1 antibody [EPR5209] (ab134113)



Western blot - Anti-Niemann Pick C1 antibody  
[EPR5209] (ab134113)

**All lanes** : Anti-Niemann Pick C1 antibody [EPR5209] (ab134113)  
at 1/10000 dilution (purified)

**Lane 1** : HepG2 (human liver hepatocellular carcinoma cell line) cell lysate

**Lane 2** : THP-1 (human monocytic leukemia cell line) cell lysate

**Lane 3** : HEK-293 (human epithelial cell line from embryonic kidney) cell lysate

**Lane 4** : PC-3 (human prostate adenocarcinoma cell line) cell lysate

Lysates/proteins at 20 µg per lane.

### Secondary

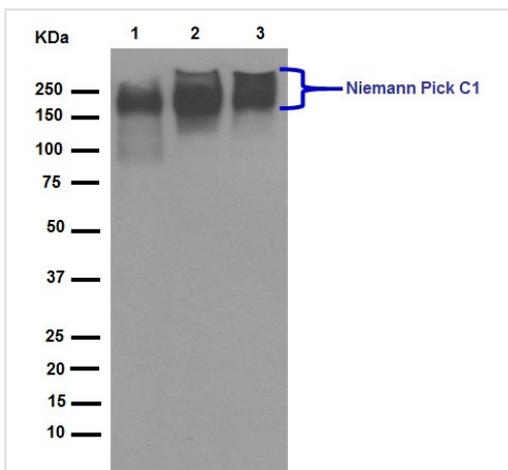
**All lanes** : HRP goat anti-rabbit (H+L) at 1/1000 dilution

**Predicted band size:** 142 kDa

**Additional bands at:** 180 kDa (possible glycosylated form)

Blocking buffer: 5% NFDM/TBST

Dilution buffer: 5% NFDM/TBST



Western blot - Anti-Niemann Pick C1 antibody  
[EPR5209] (ab134113)

**All lanes** : Anti-Niemann Pick C1 antibody [EPR5209] (ab134113)  
at 1/10000 dilution (purified)

**Lane 1** : 3T3-L1 cell lysate

**Lane 2** : L6 (rat skeletal muscle cell line) cell lysate

**Lane 3** : Rat tissue lysate

Lysates/proteins at 20 µg per lane.

### Secondary

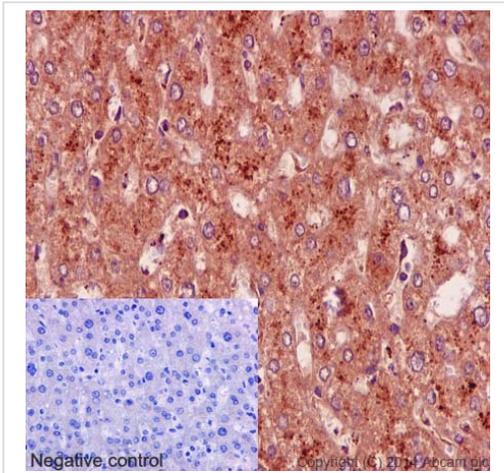
**All lanes** : HRP goat anti-rabbit (H+L) at 1/1000 dilution

**Predicted band size:** 142 kDa

**Additional bands at:** 180 kDa (possible glycosylated form)

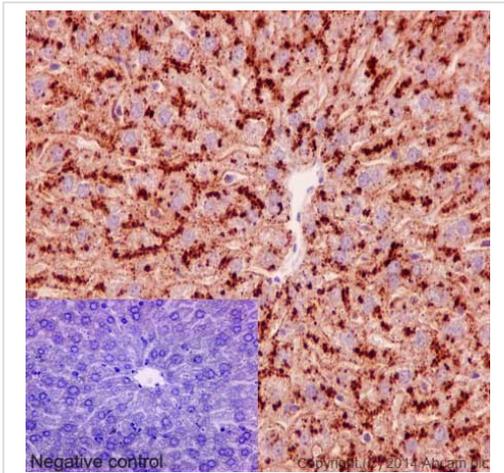
Blocking buffer: 5% NFD/MTBST

Dilution buffer: 5% NFD/MTBST



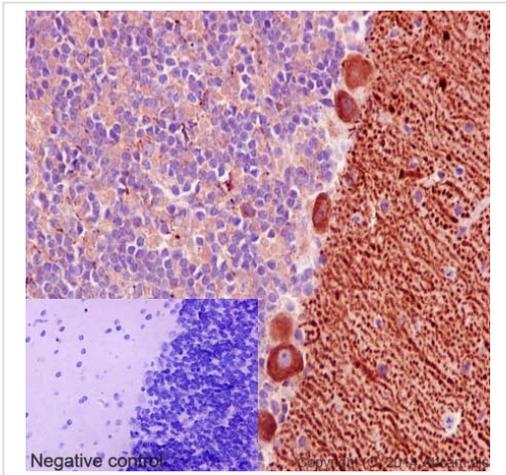
Immunohistochemical staining of paraffin embedded human liver with purified ab134113 at a working dilution of 1 in 50. The secondary antibody used is a HRP polymer for rabbit IgG. The sample is counter-stained with hematoxylin. Antigen retrieval was performed using Tris-EDTA buffer, pH 9.0. PBS was used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

Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Niemann Pick C1 antibody [EPR5209] (ab134113)



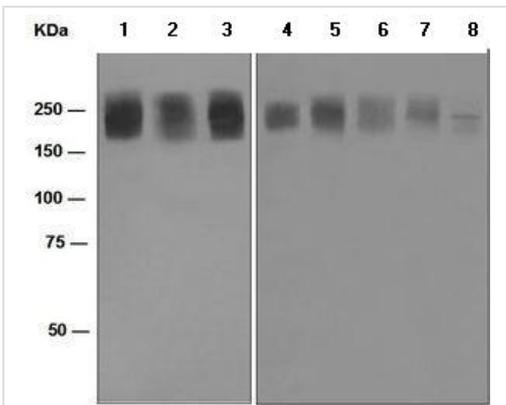
Immunohistochemical staining of paraffin embedded mouse liver with purified ab134113 at a working dilution of 1 in 50. The secondary antibody used is a HRP polymer for rabbit IgG. The sample is counter-stained with hematoxylin. Antigen retrieval was performed using Tris-EDTA buffer, pH 9.0. PBS was used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

Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Niemann Pick C1 antibody [EPR5209] (ab134113)



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 performed 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.

Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Niemann Pick C1 antibody [EPR5209] (ab134113)



Western blot - Anti-Niemann Pick C1 antibody [EPR5209] (ab134113)

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

- Lane 1** : 3T3 L1 cell lysate
- Lane 2** : L6 (rat skeletal muscle cell line) cell lysate
- Lane 3** : HepG2 (human liver hepatocellular carcinoma cell line) cell lysate
- Lane 4** : THP-1 (human monocytic leukemia cell line) cell lysate
- Lane 5** : HEK-293T (human epithelial cell line from embryonic kidney transformed with large T antigen) cell lysate
- Lane 6** : PC-3 (human prostate adenocarcinoma cell line) cell lysate
- Lane 7** : Rat liver lysate
- Lane 8** : Rat brain lysate

Lysates/proteins at 10 µg per lane.

**Secondary**

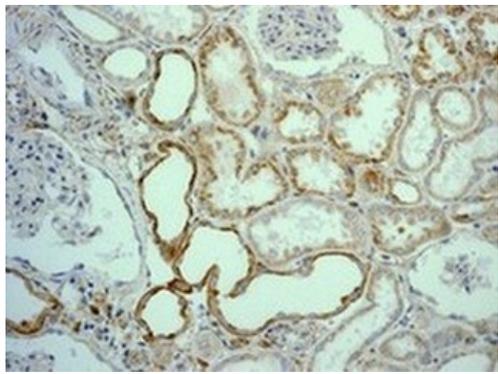
**All lanes** : Goat anti-rabbit HRP conjugated antibody at 1/2000

dilution

**Predicted band size:** 142 kDa

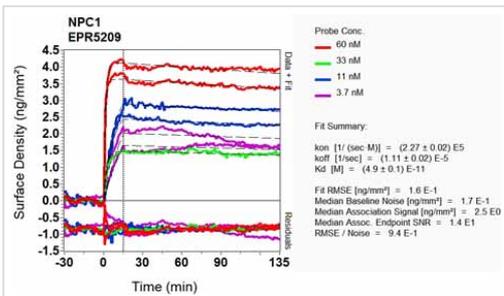
**Observed band size:** 180 kDa

[why is the actual band size different from the predicted?](#)



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

Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Niemann Pick C1 antibody [EPR5209] (ab134113)



Equilibrium dissociation constant ( $K_D$ )

[Learn more about  \$K\_D\$](#)

[Click here to learn more about  \$K\_D\$](#)

Other - Anti-Niemann Pick C1 antibody [EPR5209] (ab134113)

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

### 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