

Anti-Niemann Pick C1 antibody [EPR5209] ab134113

KO 評価済 リコンビナント RabMAb

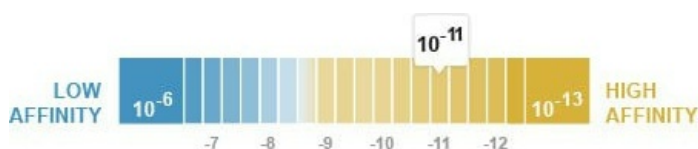
★★★★★ 2 Abreviews 42 References 画像数 14

製品の概要

製品名	Anti-Niemann Pick C1 antibody [EPR5209]
製品の詳細	Rabbit monoclonal [EPR5209] to Niemann Pick C1
由来種	Rabbit
アプリケーション	適用あり: Flow Cyt (Intra), WB, IHC-P, ICC/IF
種交差性	交差種: Mouse, Rat, Human
免疫原	Synthetic peptide within Human Niemann Pick C1 aa 1250 to the C-terminus (C terminal). The exact sequence is proprietary. Database link: O15118
ポジティブ・コントロール	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 (intra): Neuro-2a cells.
特記事項	This product is a recombinant monoclonal antibody, which offers several advantages including: <ul style="list-style-type: none"> - 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 .

製品の特性

製品の状態	Liquid
保存方法	Shipped at 4°C. Store at -20°C. Stable for 12 months at -20°C.
解離定数 (K _D 値)	K _D = 4.90 x 10 ⁻¹¹ M



[Learn more about K_D](#)

バッファー	pH: 7.2 Preservative: 0.01% Sodium azide Constituents: 40% Glycerol, 0.05% BSA, 59% PBS
精製度	Protein A purified
ポリ/モノ	モノクローナル
クローン名	EPR5209
アイソタイプ	IgG

アプリケーション

The Abpromise guarantee **Abpromise保証は、** 次のテスト済みアプリケーションにおけるab134113の使用に適用されます
アプリケーションノートには、推奨の開始希釈率がありますが、適切な希釈率につきましてはご検討ください。

アプリケーション	Abreviews	特記事項
Flow Cyt (Intra)		1/200. ab172730 - Rabbit monoclonal IgG, is suitable for use as an isotype control with this antibody.
WB	★★★★★ (1)	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)	1/70.

ターゲット情報

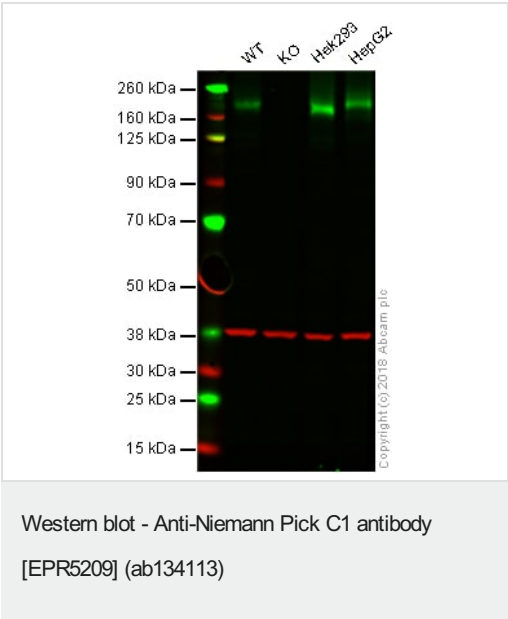
機能	Involved in the intracellular trafficking of cholesterol. May play a role in vesicular trafficking in glia, a process that may be crucial for maintaining the structural and functional integrity of nerve terminals.
関連疾患	Defects in NPC1 are the cause of Niemann-Pick disease type C1 (NPDC1) [MIM:257220]. 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.
配列類似性	Belongs to the patched family. Contains 1 SSD (sterol-sensing) domain.
ドメイン	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.

翻訳後修飾

細胞内局在

Glycosylated.
Late endosome membrane. Lysosome membrane.

画像



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

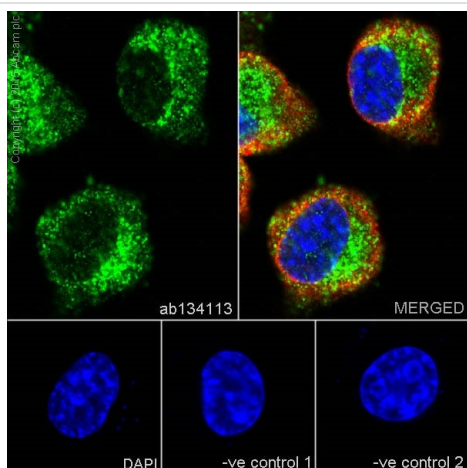
- Lane 1 :** Wild-type HAP1 whole cell lysate
- Lane 2 :** NPC1 (Niemann Pick C1) knockout HAP1 whole cell lysate
- Lane 3 :** HEK293 whole cell lysate
- Lane 4 :** HepG2 whole cell lysate

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.



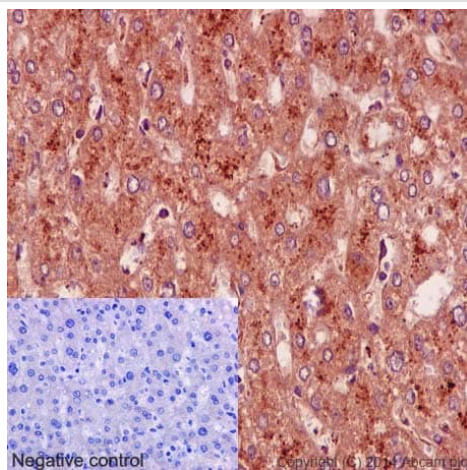
Immunocytochemistry/ Immunofluorescence - Anti-Niemann Pick C1 antibody [EPR5209] (ab134113)

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](#) (AlexaFluor®594 Goat anti-Mouse secondary) at 1/500 (red).

The negative controls are as follows:

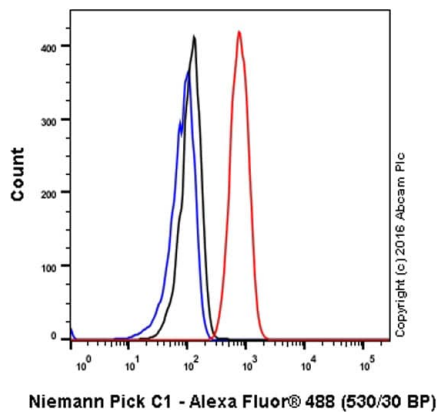
-ve control 1 – ab134113 at 1/70 followed by [ab150120](#) (AlexaFluor®594 Goat anti-Mouse secondary) at 1/500.

-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.



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

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.

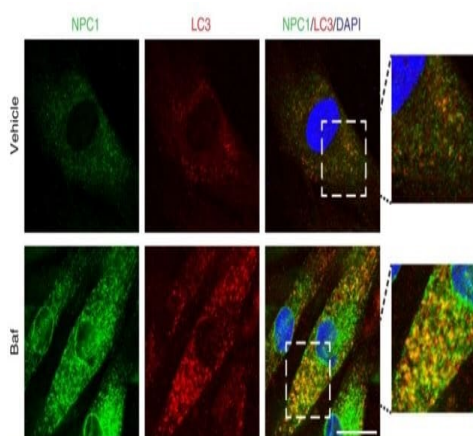


Flow Cytometry (Intracellular) - Anti-Niemann Pick C1 antibody [EPR5209] (ab134113)

ab134113 staining Niemann Pick C1 in Neuro-2a (mouse neuroblastoma cell line) by intracellular flow cytometry. Cells were fixed with 4% paraformaldehyde, permeabilised 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.

Isootype control: Rabbit monoclonal IgG (Black)

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



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

Schultz et al Nat Commun. 2018; 9: 3671. Published online 2018 Sep 10. doi: 10.1038/s41467-018-06115-2

I1061T NPC1 traffics to autophagosomes

I1061T NPC1 fibroblasts were treated with vehicle or 100 nM bafilomycin A1 (Baf) for 24 h. Fixed cells were stained for LC3 (red), NPC1 (green), and DAPI (blue) then imaged by confocal microscopy. Co-localization is indicated by yellow color in the merged image. Scale bar = 25 µm.

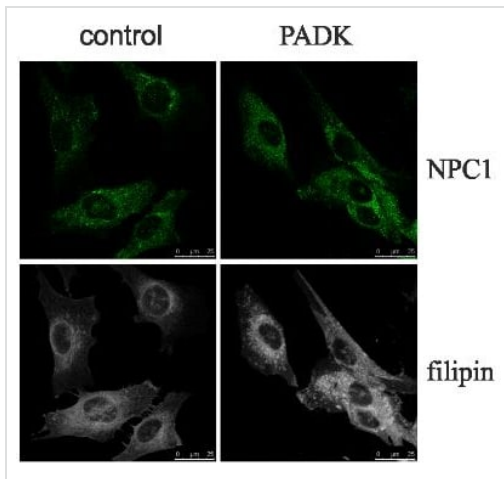
Niemann Pick C1 (NPC1) was detected using ab134113 at 1/200 dilution.

From Figure 4b of Schultz et al.

Schultz et al **Nat Commun.** 2018; 9: 3671. Published online 2018 Sep 10. doi: [10.1038/s41467-018-06115-2](https://doi.org/10.1038/s41467-018-06115-2)

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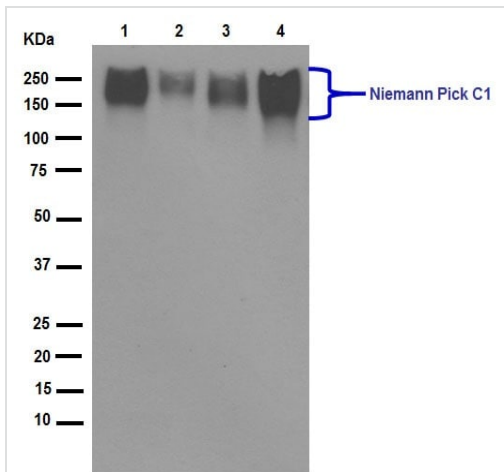


Immunocytochemistry/ Immunofluorescence - Anti-Niemann Pick C1 antibody [EPR5209] (ab134113)

Image from Cermak S et al., PLoS One. 2016;11(11):e0167428. Fig 3.; doi: 10.1371/journal.pone.0167428. Reproduced under the Creative Commons license <http://creativecommons.org/licenses/by/4.0/>

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).



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

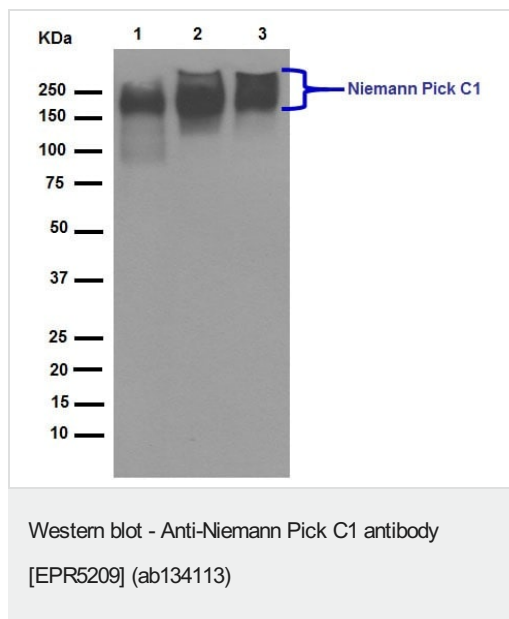
All lanes : HRP goat anti-rabbit (H+L) at 1/1000 dilution

Predicted band size: 142 kDa

Additional bands at: 180 kDa (possible glycosylated form)

Blocking buffer: 5% NFDM/TBST

Dilution buffer: 5% NFDM/TBST



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

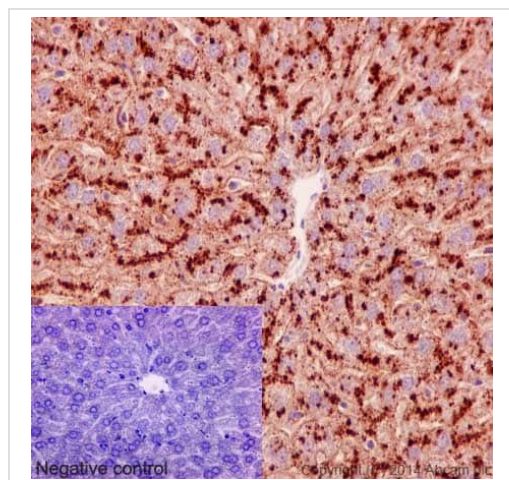
All lanes : HRP goat anti-rabbit (H+L) at 1/1000 dilution

Predicted band size: 142 kDa

Additional bands at: 180 kDa (possible glycosylated form)

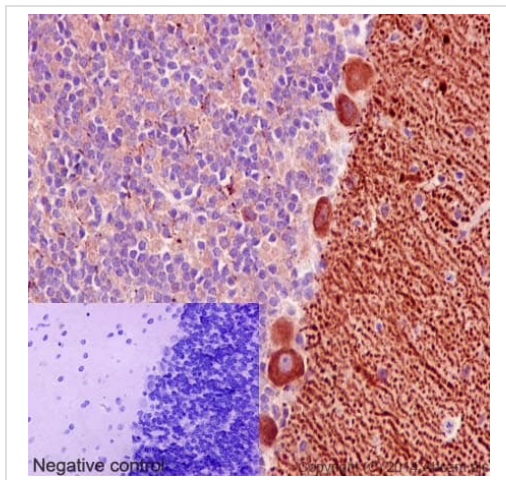
Blocking buffer: 5% NFDM/TBST

Dilution buffer: 5% NFDM/TBST



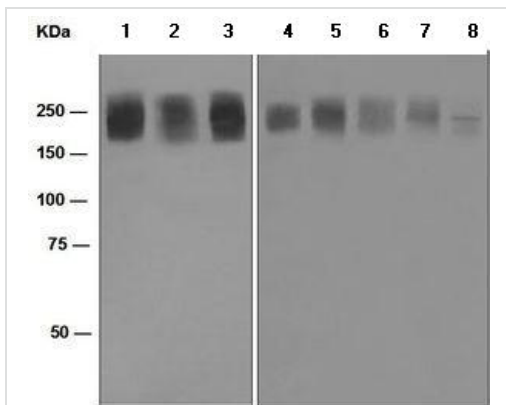
Immunohistochemical staining of paraffin embedded mouse liver with purified ab134113 at a working dilution of 1 in 50. The secondary antibody used is a HRP polymer for rabbit IgG. The sample is counter-stained with hematoxylin. Antigen retrieval was performed using Tris-EDTA buffer, pH 9.0. PBS was used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



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.



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

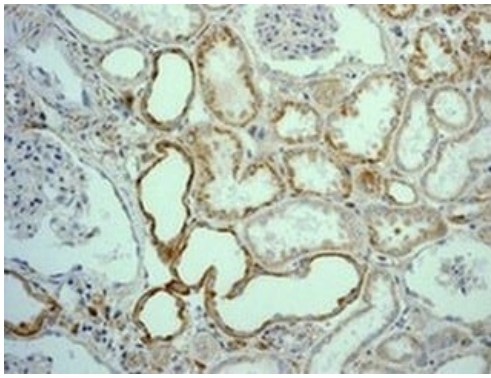
Lysates/proteins at 10 µg per lane.

Secondary

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

Predicted band size: 142 kDa

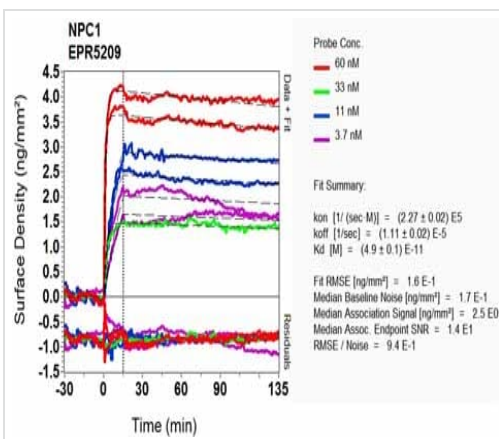
Observed band size: 180 kDa



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

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

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



OL-RD Scanning - 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\$](#)

Why choose a recombinant antibody?



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Consistent and reproducible results



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Recombinant technology



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Confirmed specificity



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Animal-free production

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

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