

DESCRIPTION

Species Reactivity	Human/Mouse/Rat
Specificity	Detects human Niemann-Pick Type C1/NPC1 in direct ELISAs. Detects human, mouse, and rat Niemann-Pick Type C1/NPC1 in Western blots.
Source	Recombinant Monoclonal Rabbit IgG ₁ Clone # 1318A
Purification	Protein A or G purified from cell culture supernatant
Immunogen	Synthetic peptide containing Human Niemann-Pick Type C1/NPC1
Formulation	Lyophilized from a 0.2 µm filtered solution in PBS with Trehalose. See Certificate of Analysis for details. *Small pack size (-SP) is supplied either lyophilized or as a 0.2 µm filtered solution in PBS.

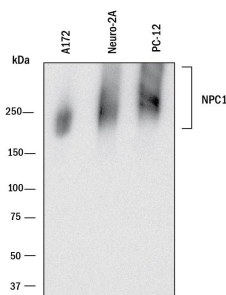
APPLICATIONS

Please Note: Optimal dilutions should be determined by each laboratory for each application. [General Protocols](#) are available in the [Technical Information](#) section on our website.

	Recommended Concentration	Sample
Western Blot	0.5 µg/mL	See Below
Immunohistochemistry	3-25 µg/mL	See Below
Knockout Validated	Niemann-Pick Type C1/NPC1 is specifically detected in HeLa human cervical epithelial carcinoma parental cell line but is not detectable in Niemann-Pick Type C1/NPC1 knockout HeLa cell line.	

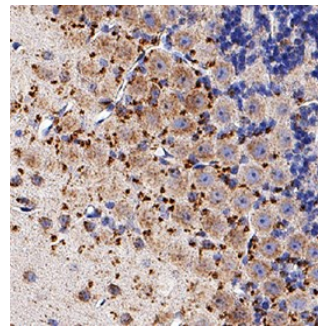
DATA

Western Blot



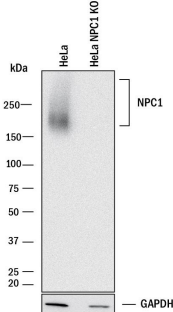
Detection of Human, Mouse, and Rat Niemann-Pick Type C1/NPC1 by Western Blot. Western blot shows lysates of A172 human glioblastoma cell line, Neuro-2A mouse neuroblastoma cell line, and PC-12 rat adrenal pheochromocytoma cell line. PVDF membrane was probed with 0.5 µg/mL of Rabbit Anti-Human/Mouse/Rat Niemann-Pick Type C1/NPC1 Monoclonal Antibody (Catalog # MAB10105) followed by HRP-conjugated Anti-Rabbit IgG Secondary Antibody (Catalog # HAF008). A specific band was detected for Niemann-Pick Type C1/NPC1 at approximately 170-250 kDa (as indicated). This experiment was conducted under reducing conditions and using [Immunoblot Buffer Group 1](#).

Immunohistochemistry



Niemann-Pick Type C1/NPC1 in Human Brain. Niemann-Pick Type C1/NPC1 was detected in immersion fixed paraffin-embedded sections of human brain (cerebellum) using Rabbit Anti-Human/Mouse/Rat Niemann-Pick Type C1/NPC1 Monoclonal Antibody (Catalog # MAB10105) at 3 µg/mL for 1 hour at room temperature followed by incubation with the Anti-Rabbit IgG VisUCyte™ HRP Polymer Antibody (Catalog # VC003). Before incubation with the primary antibody, tissue was subjected to heat-induced epitope retrieval using Antigen Retrieval Reagent-Basic (Catalog # CTS013). Tissue was stained using DAB (brown) and counterstained with hematoxylin (blue). Specific staining was localized to synaptic puncta. View our protocol for [IHC Staining with VisUCyte HRP Polymer Detection Reagents](#).

Knockout Validated



Western Blot Shows Human Niemann-Pick Type C1/NPC1 Specificity by Using Knockout Cell Line. Western blot shows lysates of HeLa human cervical epithelial carcinoma parental cell line and Niemann-Pick Type C1/NPC1 knockout HeLa cell line (KO). PVDF membrane was probed with 0.5 µg/mL of Rabbit Anti-Human/Mouse/Rat Niemann-Pick Type C1/NPC1 Monoclonal Antibody (Catalog # MAB10105) followed by HRP-conjugated Anti-Rabbit IgG Secondary Antibody (Catalog # HAF008). A specific band was detected for Niemann-Pick Type C1/NPC1 at approximately 170-250 kDa (as indicated) in the parental HeLa cell line, but is not detectable in knockout HeLa cell line. GAPDH (Catalog # MAB5718) is shown as a loading control. This experiment was conducted under reducing conditions and using [Immunoblot Buffer Group 1](#).

Human/Mouse/Rat Niemann-Pick Type C1/NPC1 Antibody

Recombinant Monoclonal Rabbit IgG₁ Clone # 1318A

Catalog Number: MAB10105

PREPARATION AND STORAGE

Reconstitution Reconstitute at 0.5 mg/mL in sterile PBS.

Shipping The product is shipped at ambient temperature. Upon receipt, store it immediately at the temperature recommended below.
*Small pack size (-SP) is shipped with polar packs. Upon receipt, store it immediately at -20 to -70 °C

Stability & Storage Use a manual defrost freezer and avoid repeated freeze-thaw cycles.

- 12 months from date of receipt, -20 to -70 °C as supplied.
- 1 month, 2 to 8 °C under sterile conditions after reconstitution.
- 6 months, -20 to -70 °C under sterile conditions after reconstitution.

BACKGROUND

NPC intracellular cholesterol transporter 1 or Niemann-Pick C1 protein (NPC1) is a 1,278 aminoacids (aa) intracellular cholesterol transporter which plays an important role in cholesterol transport from the endosomal/lysosomal compartment. NPC1 was identified as the gene that when mutated, results in Niemann-Pick disease, type C, a rare neurovisceral lipid storage disorder resulting from autosomal recessively inherited loss-of-function mutations in either NPC1 or NPC2. This disrupts intracellular lipid transport, leading to the accumulation of lipid products in the late endosomes and lysosomes. Approximately 95% of NPC patients are found to have mutations in the NPC1 gene. In humans, at least one other isoform, missing aa 519-586 is known. Human NPC1 protein sequence is 87% identical to both, mouse and rat NPC1.