abcam

Product datasheet

Anti-Niemann Pick C1 antibody ab177233

1 Image

Overview

Product name	Anti-Niemann Pick C1 antibody
Description	Goat polyclonal to Niemann Pick C1
Host species	Goat
Tested applications	**Suitable for:** WB
Species reactivity	**Reacts with:** Human
	Predicted to work with: Pig
Immunogen	Synthetic peptide corresponding to Human Niemann Pick C1 aa 1040-1053 (internal sequence) (Cysteine residue). Sequence: TYHTVLQTSADFID

Database link: **NP_000262.2**

Run BLAST with   **Run BLAST with**

Positive control	HEK293 cell lysate.
General notes	The Life Science industry has been in the grips of a reproducibility crisis for a number of years. Abcam is leading the way in addressing this with our range of recombinant monoclonal antibodies and knockout edited cell lines for gold-standard validation. Please check that this product meets your needs before purchasing.
	If you have any questions, special requirements or concerns, please send us an inquiry and/or contact our Support team ahead of purchase. Recommended alternatives for this product can be found below, along with publications, customer reviews and Q&As

Properties

Form	Liquid
Storage instructions	Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C long term. Avoid freeze / thaw cycle.
Storage buffer	pH: 7.30 Preservative: 0.02% Sodium azide Constituents: 99% Tris buffered saline, 0.5% BSA
Purity	Immunogen affinity purified

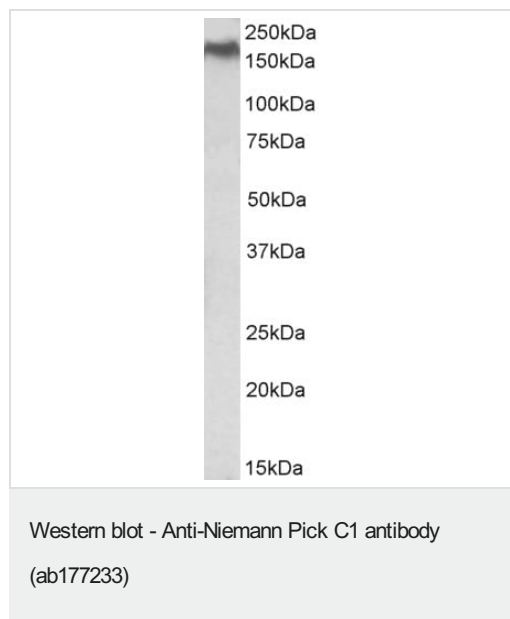| Purification notes	ab177233 was purified from goat serum by ammonium sulphate precipitation followed by antigen affinity chromatography using the immunizing peptide.
Clonality	Polyclonal
Isotype	IgG

Applications

The Abpromise guarantee   Our **Abpromise guarantee** covers the use of ab177233 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
WB		Use a concentration of 0.5 - 1.5 µg/ml. Detects a band of approximately 170 kDa (predicted molecular weight: 142 kDa). 1 hour primary incubation is recommended for this product.

Target

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

Anti-Niemann Pick C1 antibody (ab177233) at 0.5 µg/ml + HEK293 cell lysate (in RIPA buffer) at 35 µg

Developed using the ECL technique.

Predicted band size: 142 kDa
Observed band size: 170 kDa

Primary incubation was 1 hour.

Western blot - Anti-Niemann Pick C1 antibody (ab177233)

Please note: All products are "FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC PROCEDURES"

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