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NPC1 Antibody (Center)

Affinity Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP13472c

Product Information

Application	WB, IF, IHC-P, E
Primary Accession	O15118
Other Accession	NP_000262.2
Reactivity	Human, Mouse
Predicted	Rat, Bovine, Canine, Rabbit, Chicken
Host	Rabbit
Clonality	Polyclonal
Isotype	Rabbit IgG
Clone Names	RB33297
Calculated MW	142167
Antigen Region	591-620

Additional Information

Gene ID	4864
Other Names	Niemann-Pick C1 protein, NPC1
Target/Specificity	This NPC1 antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide between 591-620 amino acids from the Central region of human NPC1.
Dilution	WB~~1:2000 IF~~1:10~50 IHC-P~~1:100~500 E~~Use at an assay dependent concentration.
Format	Purified polyclonal antibody supplied in PBS with 0.05% (V/V) Proclin 300. This antibody is purified through a protein A column, followed by peptide affinity purification.
Storage	Maintain refrigerated at 2-8°C for up to 2 weeks. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.
Precautions	NPC1 Antibody (Center) is for research use only and not for use in diagnostic or therapeutic procedures.

Protein Information

Name	NPC1 (HGNC:7897)
Function	Intracellular cholesterol transporter which acts in concert with NPC2 and plays an important role in the egress of cholesterol from the

endosomal/lysosomal compartment (PubMed:10821832, PubMed:12554680, PubMed:18772377, PubMed:27238017, PubMed:9211849, PubMed:9927649). Unesterified cholesterol that has been released from LDLs in the lumen of the late endosomes/lysosomes is transferred by NPC2 to the cholesterol-binding pocket in the N-terminal domain of NPC1 (PubMed:18772377, PubMed:19563754, PubMed:27238017, PubMed:27378690, PubMed:28784760, PubMed:9211849, PubMed:9927649). Cholesterol binds to NPC1 with the hydroxyl group buried in the binding pocket (PubMed:19563754). Mediates cholesterol transfer from lysosomes to endoplasmic reticulum by tethering endosomal/lysosomal compartments and endoplasmic reticulum membrane contact sites where it interacts with sterol transport protein GRAMD1B (PubMed:31537798). Binds oxysterol with higher affinity than 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 (Probable). Inhibits cholesterol-mediated mTORC1 activation throught its interaction with SLC38A9 (PubMed:28336668).

Cellular Location Late endosome membrane; Multi-pass membrane protein. Lysosome membrane; Multi-pass membrane protein

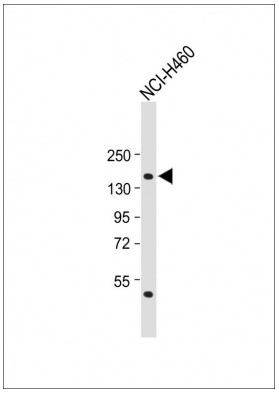
Background

This gene encodes a large protein that resides in the limiting membrane of endosomes and lysosomes and mediates intracellular cholesterol trafficking via binding of cholesterol to its N-terminal domain. It is predicted to have a cytoplasmic C-terminus, 13 transmembrane domains, and 3 large loops in the lumen of the endosome - the last loop being at the N-terminus. This protein transports low-density lipoproteins to late endosomal/lysosomal compartments where they are hydrolized and released as free cholesterol. Defects in this gene cause Niemann-Pick type C disease, a rare autosomal recessive neurodegenerative disorder characterized by over accumulation of cholesterol and glycosphingolipids in late endosomal/lysosomal compartments.

References

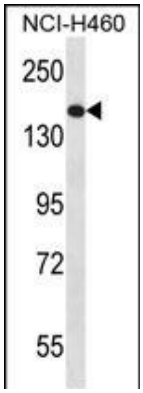
Bailey, S.D., et al. Diabetes Care 33(10):2250-2253(2010)
Fontaine-Bisson, B., et al. Diabetologia 53(10):2155-2162(2010)
Kagedal, K., et al. Biochim. Biophys. Acta 1801(8):831-838(2010)
Rodriguez-Rodriguez, E., et al. J. Alzheimers Dis. 21(2):619-625(2010)
Ma, W., et al. BMC Med. Genet. 11, 149 (2010) :

Images

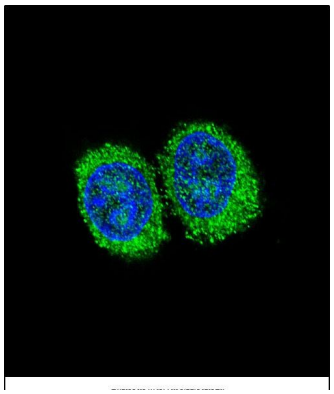


Anti-NPC1 Antibody (Center) at 1:1000 dilution + NCI-H460 whole cell lysate Lysates/proteins at 20 µg per lane. Secondary Goat Anti-Rabbit IgG, (H+L), Peroxidase conjugated at 1/10000 dilution. Predicted band size : 142 kDa Blocking/Dilution buffer: 5% NFDM/TBST.

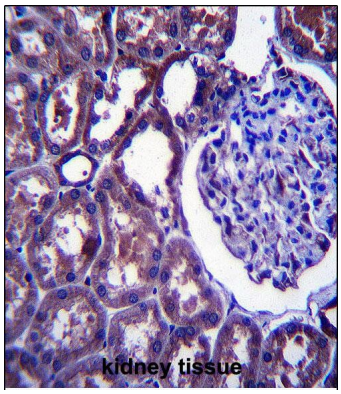
NPC1 Antibody (Center) (Cat. #AP13472c) western blot analysis in NCI-H460 cell line lysates (35ug/lane).This



demonstrates the NPC1 antibody detected the NPC1 protein (arrow).



Confocal immunofluorescent analysis of NPC1 Antibody (Center) (Cat#AP13472c) with 293 cell followed by Alexa Fluor 488-conjugated goat anti-rabbit lgG (green). DAPI was used to stain the cell nuclear (blue).



NPC1 Antibody (Center) (Cat. #AP13472c)immunohistochemistry analysis in formalin fixed and paraffin embedded human kidney tissue followed by peroxidase conjugation of the secondary antibody and DAB staining.This data demonstrates the use of NPC1 Antibody (Center) for immunohistochemistry. Clinical relevance has not been evaluated.

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