

# NPC1 Antibody (Center)

Affinity Purified Rabbit Polyclonal Antibody (Pab)

Catalog # AP13472c

## Product Information

---

<b>Application</b>	WB, IF, IHC-P, E
<b>Primary Accession</b>	<a href="#">O15118</a>
<b>Other Accession</b>	<a href="#">NP_000262.2</a>
<b>Reactivity</b>	Human, Mouse
<b>Host</b>	Rabbit
<b>Clonality</b>	Polyclonal
<b>Isotype</b>	Rabbit IgG
<b>Clone Names</b>	RB33297
<b>Calculated MW</b>	142167
<b>Antigen Region</b>	591-620

## Additional Information

---

<b>Gene ID</b>	4864
<b>Other Names</b>	Niemann-Pick C1 protein, NPC1
<b>Target/Specificity</b>	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.
<b>Dilution</b>	WB~~1:2000 IF~~1:10~50 IHC-P~~1:100~500 E~~Use at an assay dependent concentration.
<b>Format</b>	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.
<b>Storage</b>	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.
<b>Precautions</b>	NPC1 Antibody (Center) is for research use only and not for use in diagnostic or therapeutic procedures.

## Protein Information

---

<b>Name</b>	NPC1 ( <a href="#">HGNC:7897</a> )
<b>Function</b>	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: <a href="#">10821832</a> , PubMed: <a href="#">12554680</a> ,

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](#)). 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 through its interaction with SLC38A9 (PubMed:[28336668](#)).

## Cellular Location

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

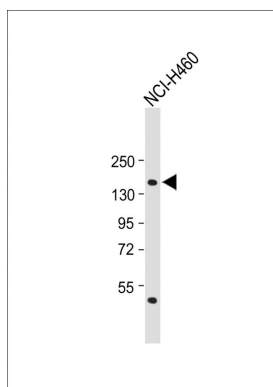
## Background

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 hydrolyzed 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

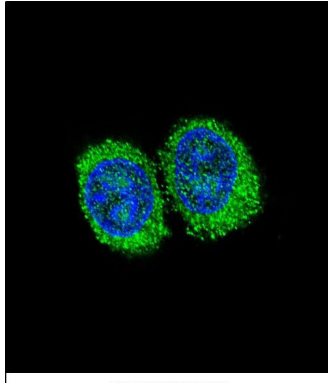
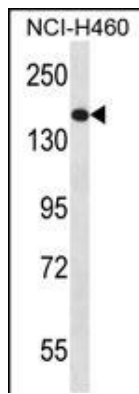
Bailey, S.D., et al. Diabetes Care 33(10):2250-2253(2010)  
Fontaine-Bisson, B., et al. Diabetologia 53(10):2155-2162(2010)  
Kagedal, K., et al. Biochim. Biophys. Acta 1801(8):831-838(2010)  
Rodriguez-Rodriguez, E., et al. J. Alzheimers Dis. 21(2):619-625(2010)  
Ma, W., et al. BMC Med. Genet. 11,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

## Images

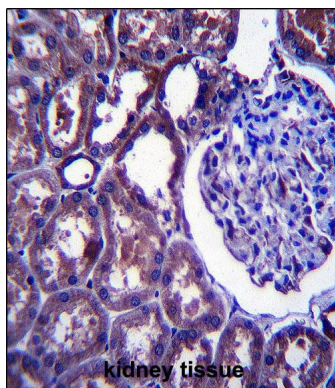


Anti-NPC1 Antibody (Center) at 1:1000 dilution + NCI-H460 whole cell lysate Lysates/proteins at 20 µg per lane. Secondary Goat Anti-Rabbit IgG, (H+L), Peroxidase conjugated at 1/10000 dilution. Predicted band size : 142 kDa Blocking/Dilution buffer: 5% NFDM/TBST.

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 IgG (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.

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.