

**NPC1 Antibody**  
Catalog # ASC10911**Specification****NPC1 Antibody - Product Information**

Application	IF
Primary Accession	<a href="#">O15118</a>
Other Accession	<a href="#">NP_000262</a> , <a href="#">4864</a>
Reactivity	Human, Mouse
Host	Rabbit
Clonality	Polyclonal
Isotype	IgG
Application Notes	NPC1 antibody can be used for detection of NPC1 by Western blot at 1 µg/mL. Antibody can also be used for immunohistochemistry starting at 2.5 µg/mL. For immunofluorescence start at 20 µg/mL.

**NPC1 Antibody - Additional Information**Gene ID **4864****Target/Specificity**

NPC1 antibody was raised against a 16 amino acid synthetic peptide from near the carboxy terminus of human NPC1. <br><br>The immunogen is located within the last 50 amino acids of NPC1.

**Reconstitution & Storage**

Antibody can be stored at 4°C up to one year. Antibodies should not be exposed to prolonged high temperatures.

**Precautions**

NPC1 Antibody is for research use only and not for use in diagnostic or therapeutic procedures.

**NPC1 Antibody - 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:<a href="http://www.uniprot.org/citations/10821832" target="\_blank">10821832</a>, PubMed:<a href="http://www.uniprot.org/citations/12554680" target="\_blank">12554680</a>, PubMed:<a href="http://www.uniprot.org/citations/18772377" target="\_blank">18772377</a>, PubMed:<a href="http://www.uniprot.org/citations/27238017" target="\_blank">27238017</a>, PubMed:<a href="http://www.uniprot.org/citations/9211849" target="\_blank">9211849</a>, PubMed:<a href="http://www.uniprot.org/citations/9927649" target="\_blank">9927649</a>). 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:<a href="http://www.uniprot.org/citations/18772377" target="\_blank">18772377</a>, PubMed:<a href="http://www.uniprot.org/citations/19563754" target="\_blank">19563754</a>, PubMed:<a href="http://www.uniprot.org/citations/27238017" target="\_blank">27238017</a>, PubMed:<a href="http://www.uniprot.org/citations/27378690" target="\_blank">27378690</a>, PubMed:<a href="http://www.uniprot.org/citations/28784760" target="\_blank">28784760</a>, PubMed:<a href="http://www.uniprot.org/citations/9211849" target="\_blank">9211849</a>, PubMed:<a href="http://www.uniprot.org/citations/9927649" target="\_blank">9927649</a>). Cholesterol binds to NPC1 with the hydroxyl group buried in the binding pocket (PubMed:<a href="http://www.uniprot.org/citations/19563754" target="\_blank">19563754</a>). 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:<a href="http://www.uniprot.org/citations/28336668" target="\_blank">28336668</a>).

### Cellular Location

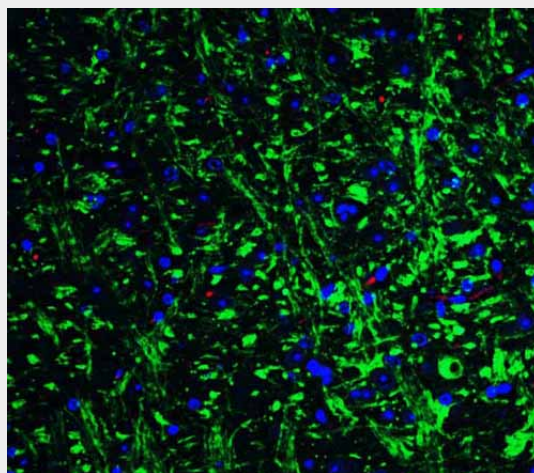
Late endosome membrane; Multi-pass membrane protein. Lysosome membrane; Multi-pass membrane protein

### NPC1 Antibody - Protocols

Provided below are standard protocols that you may find useful for product applications.

- [Western Blot](#)
- [Blocking Peptides](#)
- [Dot Blot](#)
- [Immunohistochemistry](#)
- [Immunofluorescence](#)
- [Immunoprecipitation](#)
- [Flow Cytometry](#)
- [Cell Culture](#)

### NPC1 Antibody - Images



Immunofluorescence of Grik1 in mouse brain tissue with Grik1 Antibody at 20 µg/mL.

### NPC1 Antibody - Background

NPC1 Antibody: Mutations in the Niemann-Pick disease type C1 (NPC1) gene result in a fatal

progressive neurodegenerative disorder characterized by an abnormal sequestration of lipids including cholesterol and glycosphingolipids. The NPC1 protein is 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. NPC1 transports low-density lipoproteins to late endosomal/lysosomal compartments where they are hydrolyzed and released as free cholesterol. NPC1, in addition to FTO, MC4R, and PTER has recently been shown to be a new risk loci for early-onset and morbid adult obesity in European populations. This anti-NPC1 antibody will not cross-react to NPC2, another gene whose defects also result in Niemann-Pick type C disease.

### **NPC1 Antibody - References**

Karten B, Peake KB, and Vance JE. Mechanisms and consequences of impaired lipid trafficking in Niemann-Pick type C1-deficient mammalian cells. *Biochim. Biophys. Acta*2009; 1791:656-70.  
Carstea ED, Polymeropoulos MH, Parker CC, et al. Linkage of Niemann-Pick disease type C to human chromosome 18. *Proc. Natl. Acad. Sci. USA*1993; 90:2002-4.  
Carstea ED, Morris JA, Coleman KG, et al. Niemann-Pick C1 disease gene: homology to mediators of cholesterol homeostasis. *Science*1977; 277:228-31.  
Garver WS and Heidenreich RA. The Niemann-Pick C proteins and trafficking of cholesterol through the late endosomal/lysosomal system. *Curr. Mol. Med.*2002; 2:485-505.