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NPC1 Antibody
Catalog # ASC10911

Specification

NPC1 Antibody - Product Information

Application	**WB, IHC-P, IF, E**
Primary Accession	O15118
Other Accession	NP_000262, 4864
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.

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: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). 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).

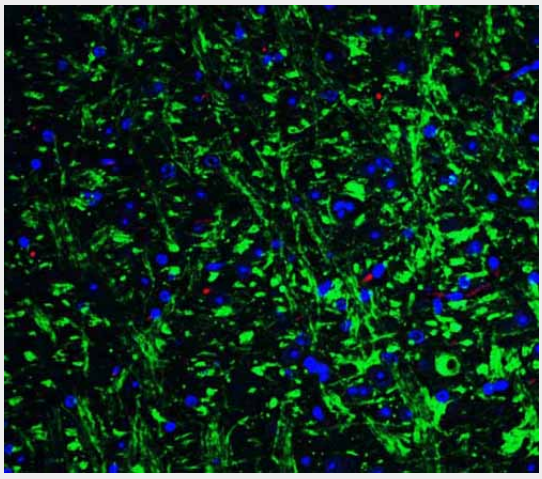
Cellular Location
Late endosome membrane; Multi-pass membrane protein. Lysosome membrane; Multi-pass membrane protein

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 Cytomety
- Cell Culture

NPC1 Antibody - Images



Immunofluorescence of Grik1 in mouse brain tissue with Grik1 Antibodyat 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 hydrolized 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. Acta2009; 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. USA1993; 90:2002-4.
Carstea ED, Morris JA, Coleman KG, et al. Niemann-Pick C1 disease gene: homology to mediators of cholesterol homeostasis. Science1977; 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.