

NPC1 Antibody

Catalog # ASC10911

Specification

NPC1 Antibody - Product Information

Application Primary Accession Other Accession Reactivity Host Clonality Isotype Application Notes IF O15118 NP_000262, 4864 Human, Mouse Rabbit Polyclonal IgG 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

Target/Specificity

4864

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:18772377, PubMed:27238017, PubMed:27238017, PubMed:9211849, PubMed:9921649, PubMed:9927649, PubMed:9927649, PubMed:9927649, PubMed:9927649, PubMed:9927649, PubMed:<a href="http://www.uniprot.org/citation



transferred by NPC2 to the cholesterol-binding pocket in the N-terminal domain of NPC1 (PubMed:18772377, PubMed:19563754, PubMed:27238017, PubMed:27378690, PubMed:27378690, PubMed:28784760, 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
- <u>Dot Blot</u>
- <u>Immunohistochemistry</u>
- Immunofluorescence
- Immunoprecipitation
- Flow Cytomety
- <u>Cell Culture</u>

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.