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NPC1 Antibody

Catalog # ASC10911

Product Information

Application	WB, IF, E, IHC-P
Primary Accession	O15118
Other Accession	NP_000262, 255652944
Reactivity	Human
Host	Rabbit
Clonality	Polyclonal
Isotype	IgG
Calculated MW	142167
Concentration (mg/ml)	1 mg/mL
Conjugate	Unconjugated
Application Notes	NPC1 antibody can be used for detection of NPC1 by Western blot at 1 g/mL. Antibody can also be used for immunohistochemistry starting at 2.5 g/mL. For immunofluorescence start at 20 g/mL.

Additional Information

Gene ID	4864
Other Names	Niemann-Pick C1 protein, NPC1
Target/Specificity	NPC1;
Reconstitution & Storage	NPC1 antibody can be stored at 4℃ for three months and -20℃, stable for up to one year. As with all antibodies care should be taken to avoid repeated freeze thaw cycles. 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.

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

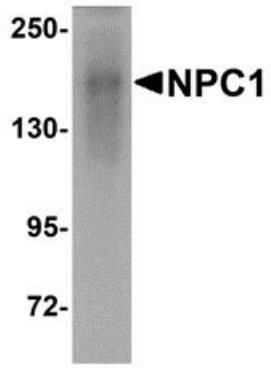
Background

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.

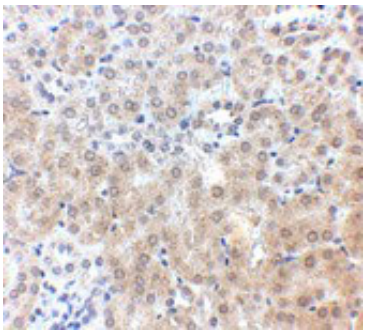
References

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.

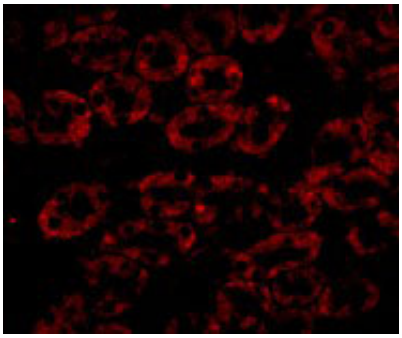
Images



Western blot analysis of NPC1 in human kidney tissue lysate with NPC1 antibody at 1 µg/mL.



Immunohistochemistry of NPC1 in mouse kidney tissue with NPC1 antibody at 2.5 µg/mL.



Immunofluorescence of NPC1 in Mouse Kidney cells with NPC1 antibody at 20 µg/mL.