

NPC1 Antibody

Purified Mouse Monoclonal Antibody Catalog # AO1840a

Product Information

Application WB, IHC, E **Primary Accession** 015118 Reactivity Human Host Mouse Clonality Monoclonal **Clone Names** 8D10B6 Isotype IgG1 **Calculated MW** 142167

Description 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 hydrolized 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.

Immunogen Purified recombinant fragment of human NPC1 (AA: 34-174) expressed in E.

Coli.

Formulation Purified antibody in PBS with 0.05% sodium azide

Additional Information

Gene ID 4864

Other Names Niemann-Pick C1 protein, NPC1

Dilution WB~~1/500 - 1/2000 IHC~~1/200 - 1/1000 E~~1/10000

Storage Maintain refrigerated at 2-8°C for up to 6 months. For long term storage store

at -20°C in small aliquots to prevent freeze-thaw cycles.

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

This gene encodes an iron containing glycoprotein which catalyzes the conversion of orthophosphoric monoester to alcohol and orthophosphate. It is the most basic of the acid phosphatases and is the only form not inhibited by L(+)-tartrate.;

References

1. Liver Int. 2010 Jul;30(6):887-97. 2. Neuroscience. 2010 May 19;167(3):608-20.

Images

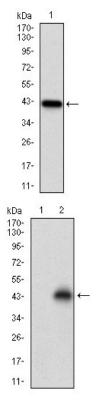


Figure 1: Western blot analysis using NPC1 mAb against human NPC1 recombinant protein. (Expected MW is 37.6 kDa)

Figure 2: Western blot analysis using NPC1 mAb against HEK293 (1) and NPC1 (AA: 34-174)-hIgGFc transfected HEK293 (2) cell lysate.

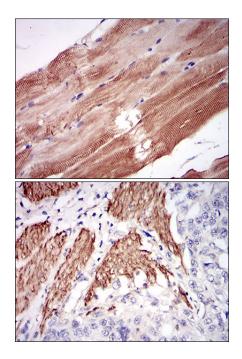


Figure 3: Immunohistochemical analysis of paraffin-embedded striated muscle tissues using NPC1 mouse mAb with DAB staining.

Figure 4: Immunohistochemical analysis of paraffin-embedded esophageal cancer tissues using NPC1 mouse mAb with DAB staining.

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