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NPC1 Antibody

Purified Mouse Monoclonal Antibody
Catalog # AO1840a

Product Information

Application	WB, IHC, E
Primary Accession	O15118
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). Mediates cholesterol transfer from lysosomes to endoplasmic reticulum by tethering endosomal/lysosomal compartments and endoplasmic reticulum membrane contact sites where it interacts with sterol transport protein GRAMD1B (PubMed:31537798). 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

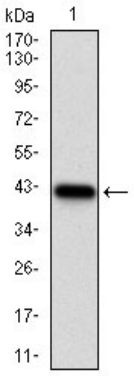
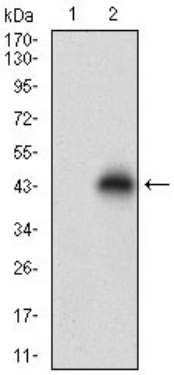


Figure 1: Western blot analysis using NPC1 mAb against human NPC1 recombinant protein. (Expected MW is 37.6 kDa)

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



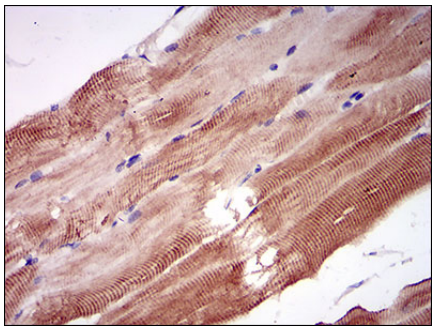


Figure 3: Immunohistochemical analysis of paraffin-embedded striated muscle tissues using NPC1 mouse mAb with DAB staining.

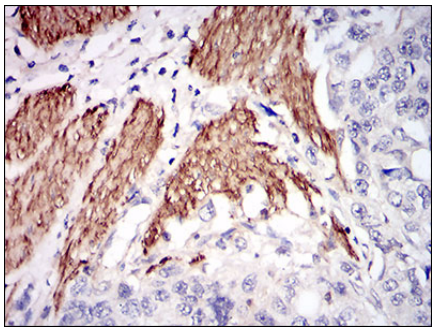


Figure 4: Immunohistochemical analysis of paraffin-embedded esophageal cancer tissues using NPC1 mouse mAb with DAB staining.

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