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Niemann Pick C2 Rabbit mAb

Catalog # AP76617

Product Information

Application	WB, IHC-P, FC
Primary Accession	P61916
Reactivity	Rat, Human, Mouse
Host	Rabbit
Clonality	Monoclonal Antibody
Isotype	IgG
Conjugate	Unconjugated
Purification	Affinity Purified
Calculated MW	16570

Additional Information

Gene ID	10577
Other Names	NPC2
Dilution	WB~~1:1000 IHC-P~~N/A FC~~1:10~50
Format	Liquid in 50mM Tris-Glycine(pH 7.4), 0.15M NaCl, 40%Glycerol, 0.01% sodium azide and 0.05% BSA.
Storage	Store at 4°C short term. Aliquot and store at -20°C long term. Avoid freeze/thaw cycles.

Protein Information

Name	NPC2 (HGNC:14537)
Synonyms	HE1
Function	Intracellular cholesterol transporter which acts in concert with NPC1 and plays an important role in the egress of cholesterol from the lysosomal compartment (PubMed:11125141, PubMed:15937921, PubMed:17018531, PubMed:18772377, PubMed:29580834). 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:17018531, PubMed:18772377, PubMed:27238017). May bind and mobilize cholesterol that is associated with membranes (PubMed:18823126). NPC2 binds cholesterol with a 1:1 stoichiometry (PubMed:17018531). Can bind a variety of sterols, including lathosterol, desmosterol and the plant sterols stigmasterol and beta-sitosterol (PubMed:17018531). The secreted form of NCP2 regulates biliary cholesterol secretion via stimulation of ABCG5/ABCG8-mediated cholesterol transport (By

	similarity).
Cellular Location	Secreted. Endoplasmic reticulum. Lysosome Note=Interaction with cell-surface M6PR mediates endocytosis and targeting to lysosomes.
Tissue Location	Detected in gallbladder bile (PubMed:21315718). Detected in fibroblasts, kidney, liver, spleen, small intestine, placenta and testis (at protein level) (PubMed:11125141). Epididymis

Background

This gene encodes a protein containing a lipid recognition domain. The encoded protein may function in regulating the transport of cholesterol through the late endosomal/lysosomal system. Mutations in this gene have been associated with Niemann-Pick disease, type C2 and frontal lobe atrophy. [provided by RefSeq, Jul 2008]