ARG59009
anti-NPC1 / Niemann Pick C1 antibody

Package: 50 µg
Store at: -20°C

Summary

Product Description	Rabbit Polyclonal antibody recognizes NPC1 / Niemann Pick C1
Tested Reactivity	Ms, Rat
Predict Reactivity	Hu
Tested Application	WB
Host	Rabbit
Clonality	Polyclonal
Isotype	IgG
Target Name	NPC1 / Niemann Pick C1
Species	Human
Immunogen	Recombinant protein corresponding to A1022-F1278 of Human NPC1 / Niemann Pick C1.
Conjugation	Un-conjugated
Alternate Names	Niemann-Pick C1 protein; NPC

Application Instructions

Application table

Application	Dilution
WB	0.1 - 0.5 µg/ml

Application Note: * The dilutions indicate recommended starting dilutions and the optimal dilutions or concentrations should be determined by the scientist.

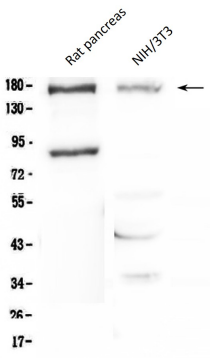
Properties

Form	Liquid
Purification	Affinity purification with immunogen.
Buffer	0.2% Na2HPO4, 0.9% NaCl, 0.05% Sodium azide and 4% Trehalose.
Preservative	0.05% Sodium azide
Stabilizer	4% Trehalose
Concentration	0.5 mg/ml
Storage instruction	For continuous use, store undiluted antibody at 2-8°C for up to a week. For long-term storage, aliquot and store at -20°C or below. Storage in frost free freezers is not recommended. Avoid repeated freeze/thaw cycles. Suggest spin the vial prior to opening. The antibody solution should be gently mixed before use.
Note	For laboratory research only, not for drug, diagnostic or other use.

Bioinformation

Gene Symbol	NPC1
Gene Full Name	Niemann-Pick disease, type C1
Background	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.[provided by RefSeq, Aug 2009]
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. Both NPC1 and NPC2 function as the cellular 'tag team duo' (TTD) to catalyze the mobilization of cholesterol within the multivesicular environment of the late endosome (LE) to effect egress through the limiting bilayer of the LE. NPC2 binds unesterified cholesterol that has been released from LDLs in the lumen of the late endosomes/lysosomes and transfers it to the cholesterol-binding pocket of the N-terminal domain of NPC1. Cholesterol binds to NPC1 with the hydroxyl group buried in the binding pocket and is exported from the limiting membrane of late endosomes/ lysosomes to the ER and plasma membrane by an unknown mechanism. 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. [UniProt]
Calculated Mw	142 kDa
PTM	Glycosylated. [UniProt]
Cellular Localization	Late endosome membrane. [UniProt]

Images



ARG59009 anti-NPC1 / Niemann Pick C1 antibody WB image

Western blot: 50 µg of samples under reducing conditions. Rat pancreas and NIH/3T3 lysates stained with ARG59009 anti-NPC1 / Niemann Pick C1 antibody at 0.5 µg/ml, overnight at 4°C.