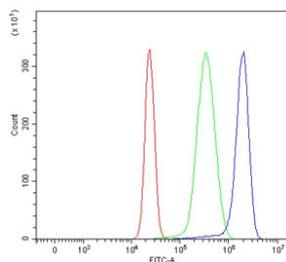


Niemann Pick C1 Antibody / NPC1 (RQ7282)

Catalog No.	Formulation	Size
RQ7282	0.5mg/ml if reconstituted with 0.2ml sterile DI water	100 ug

[Bulk quote request](#)

Availability	1-3 business days
Species Reactivity	Human, Mouse, Rat
Format	Antigen affinity purified
Host	Rabbit
Clonality	Polyclonal (rabbit origin)
Isotype	Rabbit IgG
Purity	Antigen affinity purified
Buffer	Lyophilized from 1X PBS with 2% Trehalose
UniProt	O15118
Applications	Western Blot : 0.5-1ug/ml Flow Cytometry : 1-3ug/million cells
Limitations	This Niemann Pick C1 antibody is available for research use only.



Flow cytometry testing of human HepG2 cells with Niemann Pick C1 antibody at 1ug/million cells (blocked with goat sera); Red=cells alone, Green=isotype control, Blue=Niemann Pick C1 antibody.



Western blot testing of 1) human 293T, 2) human HepG2, 3) human HeLa and 4) rat PC-12 cell lysate with Niemann Pick C1 antibody. Predicted molecular weight ~142/170~190 kDa (unmodified/glycosylated).

Description

Niemann-Pick disease, type C1 (NPC1) is a membrane protein that mediates intracellular cholesterol trafficking in mammals. In humans it is encoded by the NPC1 gene (chromosome location 18q11). 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-absorbance lipoproteins to late endosomal/lysosomal compartments where they are hydrolyzed 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.

Application Notes

Optimal dilution of the Niemann Pick C1 antibody should be determined by the researcher.

Immunogen

Amino acids YHTVLQTSADFIDALKKARLI were used as the immunogen for the Niemann Pick C1 antibody.

Storage

After reconstitution, the Niemann Pick C1 antibody can be stored for up to one month at 4°C. For long-term, aliquot and store at -20°C. Avoid repeated freezing and thawing.