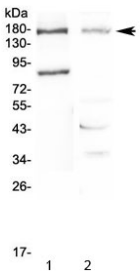
NPC1 Antibody / Niemann Pick C1 (RQ4395)

Catalog No.	Formulation	Size
RQ4395	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 and 0.025% sodium azide
UniProt	O15118
Applications	Western blot: 0.5-1ug/ml Direct ELISA: 0.1-0.5ug/ml (recombinant human protein) (BSA-free format available)
Limitations	This NPC1 antibody is available for research use only.



Western blot testing of 1) rat pancreas and 2) mouse NIH 3T3 lysate with NPC1 antibody at 0.5ug/ml. Expected molecular weight: 140-190 kDa depending on glycosylation level.

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-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.

Application Notes

Optimal dilution of the NPC1 antibody should be determined by the researcher.

Immunogen

A human partial recombinant protein corresponding to amino acids A1022-F1278 was used as the immunogen for the NPC1 antibody.

Storage

After reconstitution, the NPC1 antibody can be stored for up to one month at 4oC. For long-term, aliquot and store at -20oC. Avoid repeated freezing and thawing.

Copyright © NSJ Bioreagents. All rights reserved.