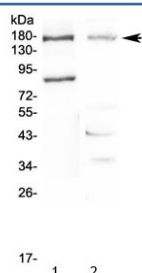


## NPC1 Antibody / Niemann Pick C1 (RQ4395)

Catalog No.	Formulation	Size
RQ4395	0.5mg/ml if reconstituted with 0.2ml sterile DI water	100 ug

**Bulk quote request**

<b>Availability</b>	1-3 business days
<b>Species Reactivity</b>	Human, Mouse, Rat
<b>Format</b>	Antigen affinity purified
<b>Clonality</b>	Polyclonal (rabbit origin)
<b>Isotype</b>	Rabbit IgG
<b>Purity</b>	Antigen affinity purified
<b>Buffer</b>	Lyophilized from 1X PBS with 2% Trehalose and 0.025% sodium azide
<b>UniProt</b>	O15118
<b>Applications</b>	Western Blot : 0.5-1ug/ml Direct ELISA : 0.1-0.5ug/ml (recombinant human protein) (BSA-free format available)
<b>Limitations</b>	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 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 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 4°C. For long-term, aliquot and store at -20°C. Avoid repeated freezing and thawing.