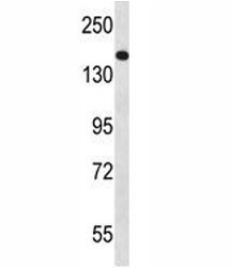
NPC1 Antibody / Niemann Pick C1 (F42992)

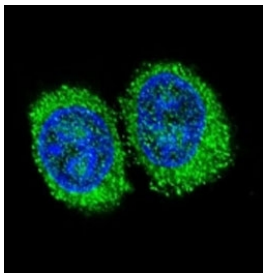
Catalog No.	Formulation	Size
F42992-0.4ML	In 1X PBS, pH 7.4, with 0.09% sodium azide	0.4 ml
F42992-0.08ML	In 1X PBS, pH 7.4, with 0.09% sodium azide	0.08 ml

Bulk quote request

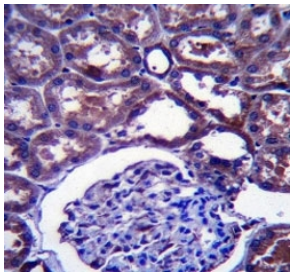
Availability	1-3 business days
Species Reactivity	Human
Format	Antigen affinity purified
Host	Rabbit
Clonality	Polyclonal (rabbit origin)
Isotype	Rabbit Ig
Purity	Antigen affinity
UniProt	O15118
Applications	Western Blot : 1:1000 Immunofluorescence : 1:10-1:50 Immunohistochemistry (Paraffin) : 1:10-1:50
Limitations	This NPC1 antibody is available for research use only.



NPC1 antibody western blot analysis in NCI-H460 lysate. Predicted molecular weight ~142/170~190 kDa (unmodified/glycosylated).



Confocal immunofluorescent analysis of NPC1 antibody with 293 cells followed by Alexa Fluor 488-conjugated goat anti-rabbit IgG (green). DAPI was used as a nuclear counterstain (blue).



NPC1 antibody immunohistochemistry analysis in formalin fixed and paraffin embedded human kidney tissue.

Description

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

Titration of the NPC1 antibody may be required due to differences in protocols and secondary/substrate sensitivity.

Immunogen

A portion of amino acids 591-620 from the human protein was used as the immunogen for this NPC1 antibody.

Storage

Aliquot the NPC1 antibody and store frozen at -20oC or colder. Avoid repeated freeze-thaw cycles.