

For Research Use Only

NPC1 Recombinant monoclonal antibody

Catalog Number:86233-5-RR



Basic Information

Catalog Number: 86233-5-RR	GenBank Accession Number: BC063302	Purification Method: Protein A purification
Source: Rabbit	GeneID (NCBI): 4864	CloneNo.: 250854D2
Isotype: IgG	UNIPROT ID: O15118	Recommended Dilutions: WB: 1:2000-1:10000 IF/ICC: 1:200-1:800
Immunogen Catalog Number: AG4946	Full Name: Niemann-Pick disease, type C1	
	Calculated MW: 142 kDa	
	Observed MW: 160-200 kDa	

Applications

Tested Applications: WB, IF/ICC, ELISA	Positive Controls: WB : 37°C incubated HEK-293 cells, IF/ICC : HepG2 cells,
Species Specificity: human	

Background Information

Niemann-Pick Type C (NPC) disease is a lysosomal storage disorder characterized by the accumulation of unesterified cholesterol and other lipids in the endolysosomal system. NPC disease results from a defect in either of two distinct cholesterol-binding proteins: a transmembrane protein, NPC1, and a small soluble protein, NPC2. NPC1 or NPC2 deficiency models showed that the functions of these two proteins within lysosomes are linked closely. NPC1 is a typical transmembrane protein and contains a number of modification sites for glycosylation. Defects in NPC1 are the cause of Niemann-Pick disease type C1 which exhibits highly variable clinical phenotype. Moreover, NPC1 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.

Storage

Storage:
Store at -20°C. Stable for one year after shipment.
Storage Buffer:
PBS with 0.02% sodium azide and 50% glycerol, pH7.3
Aliquoting is unnecessary for -20°C storage

For technical support and original validation data for this product please contact:

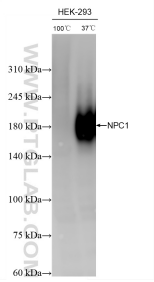
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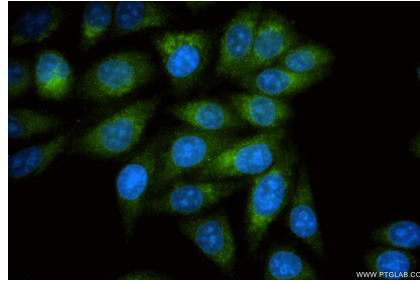
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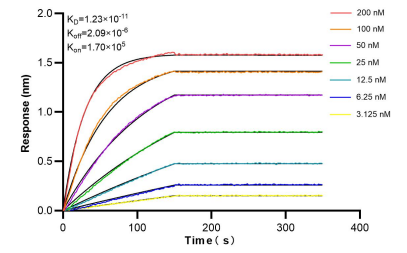
Selected Validation Data



100°C incubated and 37°C incubated HEK-293 cell lysates were subjected to SDS PAGE followed by western blot with 86233-5-RR (NPC1 antibody) at dilution of 1:5000 incubated at room temperature for 1.5 hours.



Immunofluorescent analysis of (-20°C Methanol) fixed HepG2 cells using NPC1 antibody (86233-5-RR, Clone: 250854D2) at dilution of 1:400 and CoraLite® 488-Conjugated Goat Anti-Rabbit IgG(H+L) (SA00013-2).



Bi-layer interferometry (BLI) kinetic assays of 86233-5-RR against Human NPC1 were performed. The affinity constant is 12.3 pM.