Product Datasheet

Niemann Pick C1 Rabbit mAb

Catalog No: #49838

Package Size: #49838-1 50ul #49838-2 100ul

Orders: order@signalwayantibody.com
Support: tech@signalwayantibody.com

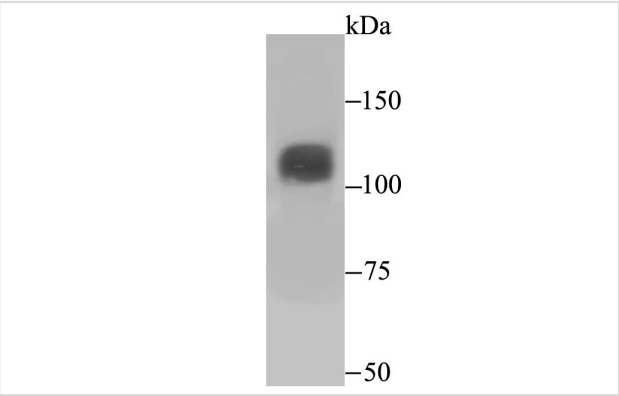
Description

Product Name	Niemann Pick C1 Rabbit mAb
Host Species	Recombinant Rabbit
Clonality	Monoclonal antibody
Clone No.	JB87-33
Purification	ProA affinity purified
Applications	WB,ICC,IF,IHC,FC
Species Reactivity	Hu, Ms, Rt
Immunogen Description	Recombinant protein
Other Names	Niemann Pick C1 protein precursor antibody Niemann Pick disease, type C1 antibody Niemann-Pick C1 protein antibody NPC antibody NPC1 antibody NPC1_HUMAN antibody
Accession No.	Swiss-Prot#:O15118
Uniprot	O15118
GeneID	4864;
Calculated MW	142 kDa
Formulation	1*TBS (pH7.4), 1%BSA, 40%Glycerol. Preservative: 0.05% Sodium Azide.
Storage	Store at -20°C

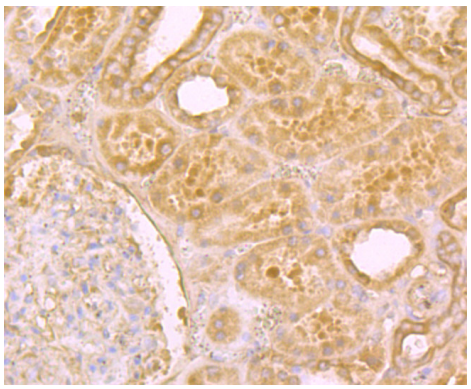
Application Details

WB: 1:500-1:1,000 IHC: 1:50-1:200 ICC/IF: 1:50-1:200FC: 1:50-1:100

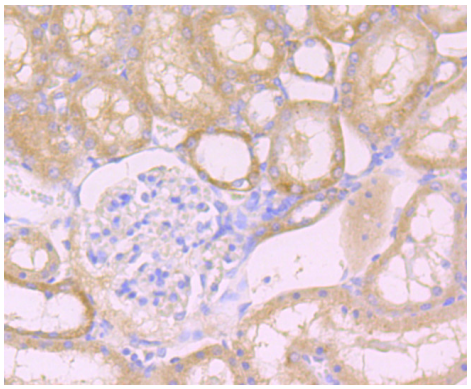
Images



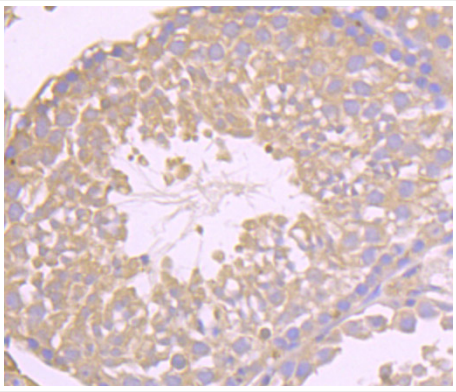
Western blot analysis of Niemann Pick C1 on SiHa cell lysate using anti-Niemann Pick C1 antibody at 1/500 dilution.



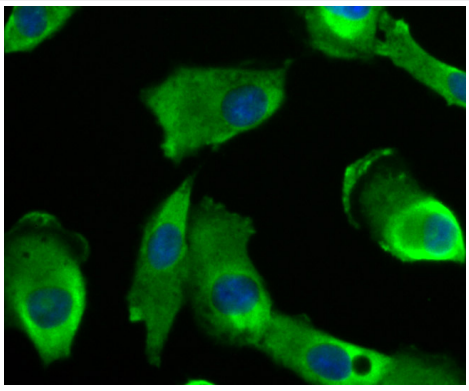
Immunohistochemical analysis of paraffin-embedded human kidney tissue using anti-Niemann Pick C1 antibody. Counter stained with hematoxylin.



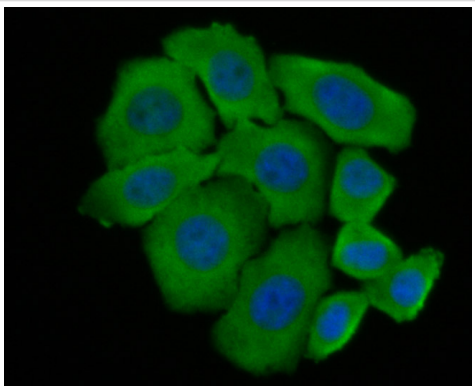
Immunohistochemical analysis of paraffin-embedded rat kidney tissue using anti-Niemann Pick C1 antibody. Counter stained with hematoxylin.



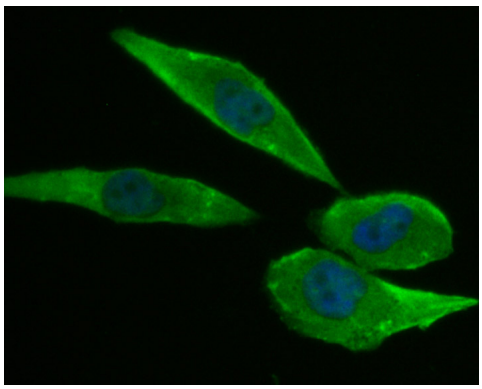
Immunohistochemical analysis of paraffin-embedded mouse testis tissue using anti-Niemann Pick C1 antibody. Counter stained with hematoxylin.



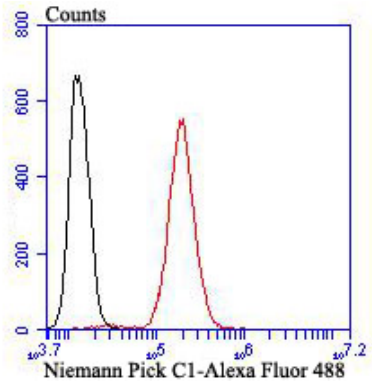
ICC staining Niemann Pick C1 in A549 cells (green). The nuclear counter stain is DAPI (blue). Cells were fixed in paraformaldehyde, permeabilised with 0.25% Triton X100/PBS.



ICC staining Niemann Pick C1 in HepG2 cells (green). The nuclear counter stain is DAPI (blue). Cells were fixed in paraformaldehyde, permeabilised with 0.25% Triton X100/PBS.



ICC staining Niemann Pick C1 in PC-3M cells (green). The nuclear counter stain is DAPI (blue). Cells were fixed in paraformaldehyde, permeabilised with 0.25% Triton X100/PBS.



Flow cytometric analysis of SH-SY-5Y cells with Niemann Pick C1 antibody at 1/100 dilution (red) compared with an unlabelled control (cells without incubation with primary antibody; black). Alexa Fluor 488-conjugated goat anti rabbit IgG was used as the secondary antibody.

Background

Cells obtain cholesterol via two distinct pathways, endogenous synthesis in the endoplasmic reticulum and exogenous uptake through the low-density lipoprotein (LDL) receptor pathway. NPC1 is a protein that resides in late endosomes and lysosomes and is involved in the intracellular trafficking of cholesterol. The human NPC1 gene maps to chromosome 18q11.2 and produces proteins which undergo N-glycosylation and are expressed in brain and liver. NPC1 contains a cysteine-rich domain, which is critical for proper protein function, but is highly mutated. Mutations in NPC1 result in Niemann-Pick disease type C (NPC), an autosomal recessive disease characterized by the accumulation of unesterified cholesterol in the endosomal/lysosomal system. The accumulation of cholesterol results in progressive neurodegeneration and death. More than 90% of cases of NPC are due to mutations in NPC1 and patients with NPC display multiple neurological symptoms, such as hepatosplenomegaly, ataxia, dystonia and dementia.

References

Note: This product is for in vitro research use only