Data Sheet (Cat.No.TMAC-02848)

Anti-Niemann Pick C1/NPC1 Antibody (1B891)

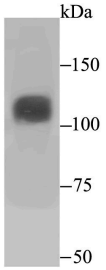
Product Details

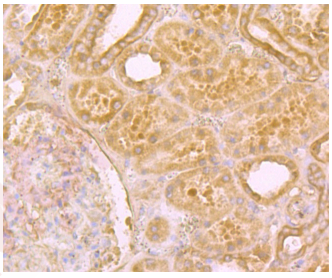
Ig Type:	IgG
Reactivity:	Human,Mouse,Rat
Conjugation:	Unconjugated
Molecular Weight:	Theoretical: 142 kDa.
Clone:	1B891
Purification:	ProA affinity purified

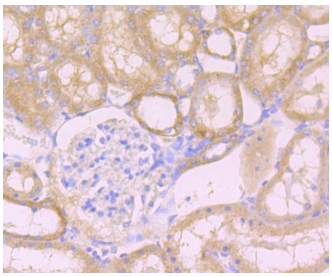
Applications

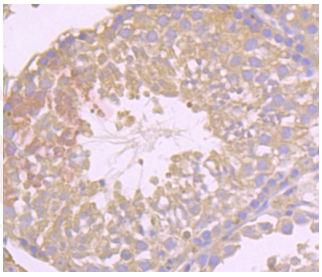
Verified Activity:

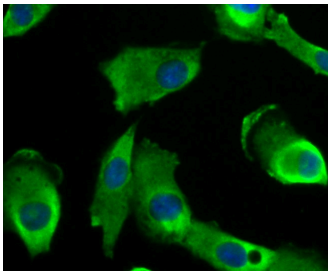
1. Western blot analysis of Niemann Pick C1 on SiHa cell lysate using anti-Niemann Pick C1 antibody at 1/500 dilution.
2. Immunohistochemical analysis of paraffin-embedded human kidney tissue using anti-Niemann Pick C1 antibody. Counter stained with hematoxylin.
3. Immunohistochemical analysis of paraffin-embedded rat kidney tissue using anti-Niemann Pick C1 antibody. Counter stained with hematoxylin.
4. Immunohistochemical analysis of paraffin-embedded mouse testis tissue using anti-Niemann Pick C1 antibody. Counter stained with hematoxylin.
5. 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.
6. 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.
7. 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.
8. 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.

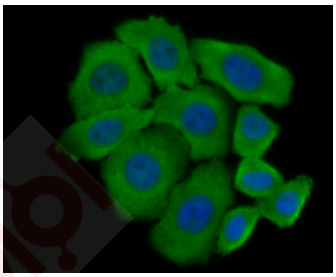


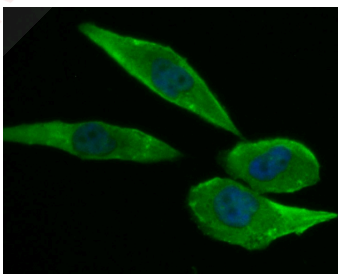


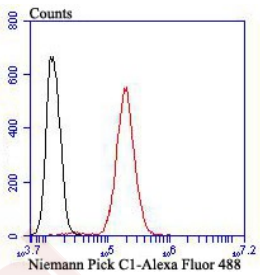












Application: FCM,ICC,IF,IHC,WB

Recommended: WB: 1:500-1000; IHC: 1:50-200; ICC:IF: 1:50-200; FCM: 1:50-100

Properties

Stability & Storage: Store at -20°C or -80°C for 12 months. Avoid repeated freeze-thaw cycles.

Shipping: Shipping with blue ice.

Antigen Details

Immunogen: Recombinant Protein

Uniprot ID: O15118

Synonyms: NPC;Niemann-Pick disease, type C1

Research 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.

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