

Niemann Pick C1 polyclonal antibody

Catalog: BS90945

Host: Rabbit

Reactivity: Human, Mouse, Rat

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.

Product:

Rabbit IgG, 1mg/ml in PBS with 0.02% sodium azide, 50% glycerol, pH7.2

Molecular Weight:

142 kDa

Swiss-Prot:

O15118(Human) O35604(Mouse)

Purification&Purity:

ProA affinity purified

Applications:

WB:1:500-1:1,000

ICC/IF:1:50-1:200

IHC:1:50-1:200

FC:1:50-1:100

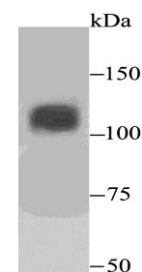
Storage&Stability:

Store at +4 °C after thawing. Aliquot store at -20 °C or -80 °C. Avoid repeated freeze / thaw cycles.

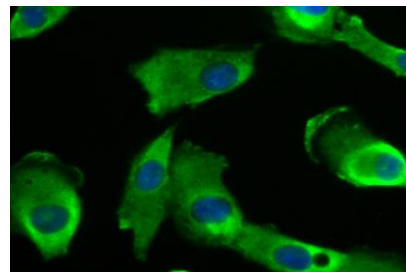
Specificity:

Niemann Pick C1 polyclonal antibody detects endogenous levels of Niemann Pick C1 protein.

DATA:



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



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.

Note:

For research use only, not for use in diagnostic procedure.

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