Niemann Pick C1 Recombinant Rabbit mAb

Catalog: BS48092 Host: Rabbit Reactivity: Human, Mouse, Rat

BackGround:

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.[provided by RefSeq, Aug 2009]

Product:

Store at -20℃. Supplied in 50mM Tris-Glycine(pH 7.4), 0.15M NaCl, 40%Glycerol, 0.01% sodium azide and 0.05% BSA. Stable for 12 months from date of receipt.

Molecular Weight:

160-180 kDa

Swiss-Prot:

O15118

Purification&Purity:

Affinity Purification

Applications:

WB: 1:2000
IHC: 1:100
ICC/IF: 1:100
FC: 1:50

Storage&Stability:

Store at 4℃ short term. Aliquot and store at -20℃ long term. Avoid freeze-thaw cycles.

Isotype:

IgG

DATA:

Note:

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

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