abcam

Product datasheet

Anti-Niemann Pick C1 antibody ab36983

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概述

产品名称	Anti-Niemann Pick C1抗体
描述	**兔多克隆抗体**to Niemann Pick C1
宿主	Rabbit
特异性	ab36983 recognises heterogeneously glycosylated Niemann Pick C1 protein.
经测试应用	**适用于:** Electron Microscopy, WB, ICC/IF, IP
种属反应性	**与反应:** Rat, Hamster, Human **预测可用于:** Mouse
免疫原	Synthetic peptide made to the C terminal region of human Niemann Pick C1.
阳性对照	Human fibroblast cell lysate
常规说明	The mouse reactivity has been predicted based on the positive data published in publications e.g. PMID 21856732, PMID20007718, PMID18815434, PMID18848945 etc.

性能

形式	Liquid
存放说明	Shipped at 4°C. Upon delivery aliquot and store at -20°C. Avoid freeze / thaw cycles.
存储溶液	Preservative: 0.025% Sodium azide Constituent: PBS
纯度	Protein A purified
克隆	**多克隆**
同种型	IgG

应用

Our Abpromise guarantee covers the use of **ab36983** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

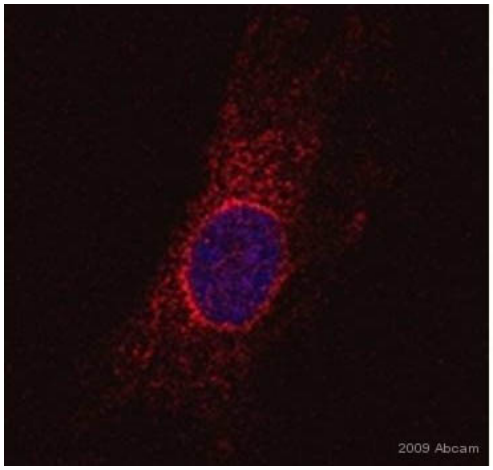
应用	Ab评论	说明
Electron Microscopy		Use at an assay dependent dilution.

应用	Ab评论	说明
WB	★★★★★	1/1000 - 1/3000. Predicted molecular weight: 142 kDa.
ICC/IF	★★★★★	1/250.
IP		Use at an assay dependent dilution.

靶标

功能	Involved in the intracellular trafficking of cholesterol. 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.
疾病相关	Defects in NPC1 are the cause of Niemann-Pick disease type C1 (NPDC1) [MIM:257220]. A lysosomal storage disorder that affects the viscera and the central nervous system. It is due to defective intracellular processing and transport of low-density lipoprotein derived cholesterol. It causes accumulation of cholesterol in lysosomes, with delayed induction of cholesterol homeostatic reactions. Niemann-Pick disease type C1 has a highly variable clinical phenotype. Clinical features include variable hepatosplenomegaly and severe progressive neurological dysfunction such as ataxia, dystonia and dementia. The age of onset can vary from infancy to late adulthood. An allelic variant of Niemann-Pick disease type C1 is found in people with Nova Scotia ancestry. Patients with the Nova Scotian clinical variant are less severely affected.
序列相似性	Belongs to the patched family. Contains 1 SSD (sterol-sensing) domain.
结构域	A cysteine-rich N-terminal domain and a C-terminal domain containing a di-leucine motif necessary for lysosomal targeting are critical for mobilization of cholesterol from lysosomes.
翻译后修饰	Glycosylated.
细胞定位	Late endosome membrane. Lysosome membrane.

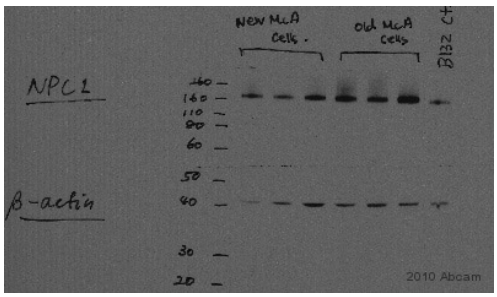
图片



Immunocytochemistry/ Immunofluorescence - Anti-Niemann Pick C1 antibody (ab36983)

This image was kindly supplied by Daniel Rodriguez by Abreview

ab36983 at a 1/800 dilution staining Niemann Pick C1 in mouse 3T3-L1 cells by Immunocytochemistry/ Immunofluorescence, incubated for 12 hours at 4°C. Fixed in methanol. Blocked with 1% BSA for 1 hour 30 minutes at room temperature. Secondary used at 1/800 dilution monoclonal Goat anti-rabbit conjugated to Alexa Fluor 568 (red). Nucleus was stained with DAPI (blue).
Note localization of Niemann Pick C1 around the nucleus and in cytosolic vesicles.



Western blot - Anti-Niemann Pick C1 antibody (ab36983)

This image is courtesy of an anonymous Abreview

All lanes : Niemann Pick C1 antibody (ab36983) at 1/1000 (for 12 hours at 4°C) + whole cell lysate of Rat Hepatoma at 30 µg

Secondary
An undiluted HRP-conjugated Goat anti-rabbit polyclonal developed using the ECL technique

Performed under reducing conditions.

Exposure time ; 1 minute

Blocking Step : 5% Milk for 1 hour at 25°C

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