abcam

Product datasheet

Recombinant Human Niemann Pick C1 protein ab114306

画像数 1

製品の概要

製品名	Recombinant Human Niemann Pick C1 protein
タンパク質長	Protein fragment

製品の詳細

由来	Recombinant
由来	Wheat germ
アミノ酸配列	
アクセッション番号	O15118
生物種	Human
配列	GFANAMYNACRDVEAPSSNDKALGLLCGKDADACNATNWIEYMFNKDNGQ APFTITPVFSDFPVHGMEPMNNATKGCDESVDEVTAPCSCQDCSIVCGPK
分子量	37 kDa including tags
領域	151 to 250

特性

Our Abpromise guarantee covers the use of **ab114306** in the following tested applications.

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

アプリケーション	Western blot SDS-PAGE ELISA
製品の状態	Liquid
備考	Protein concentration is above or equal to 0.05 mg/ml. This protein is best used within three months from the date of receipt.

前処理および保存

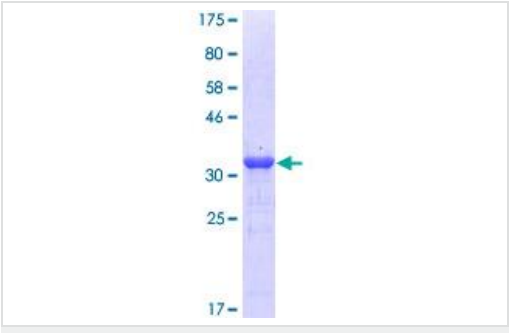
保存方法および安定性	Shipped on dry ice. Upon delivery aliquot and store at -80ºC. Avoid freeze / thaw cycles. pH: 8.00

Constituents: 0.3% Glutathione, 0.79% Tris HCl

関連情報

機能	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.
関連疾患	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.
配列類似性	Belongs to the patched family. Contains 1 SSD (sterol-sensing) domain.
ドメイン	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.
翻訳後修飾	Glycosylated.
細胞内局在	Late endosome membrane. Lysosome membrane.

画像



SDS-PAGE - Recombinant Human Niemann Pick C1 protein (ab114306)

12.5% SDS-PAGE image showing ab114306
Stained with Coomassie Blue.

Our Abpromise to you: Quality guaranteed and expert technical support

- Replacement or refund for products not performing as stated on the datasheet
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- Response to your inquiry within 24 hours

- We provide support in Chinese, English, French, German, Japanese and Spanish
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- We investigate all quality concerns to ensure our products perform to the highest standards

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