SANTA CRUZ BIOTECHNOLOGY, INC.

NPC1 (C-21): sc-18201

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

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 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.

REFERENCES

1. Watari, H., et al. 1999. Mutations in the leucine zipper motif and sterol-sensing domain inactivate the Niemann-Pick C1 glycoprotein. J. Biol. Chem. 274: 21861-21866.
2. Greer, W.L., et al. 1999. Mutations in NPC1 highlight a conserved NPC1-specific cysteine-rich domain. Am. J. Hum. Genet. 65: 1252-1260.
3. Sym, M., et al. 2000. A model for niemann-pick type C disease in the nematode *Caenorhabditis elegans*. Curr. Biol. 10: 527-530.
4. Cruz, J.C., et al. 2000. Fate of endogenously synthesized cholesterol in Niemann-Pick type C1 cells. J. Biol. Chem. 275: 41309-41316.
5. Sun, X., et al. 2001. Niemann-Pick C variant detection by altered sphingolipid trafficking and correlation with mutations within a specific domain of NPC1. Am. J. Hum. Genet. 68: 1361-1372.
6. Watabe, K., et al. 2001. Establishment and characterization of immortalized Schwann cells from murine model of Niemann-Pick disease type C (spm/spm). J. Peripher. Nerv. Syst. 6: 85-94.

CHROMOSOMAL LOCATION

Genetic locus: NPC1 (human) mapping to 18q11.2; Npc1 (mouse) mapping to 18 A1.

SOURCE

NPC1 (C-21) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the C-terminus of NPC1 of human origin.

PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-18201 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

RESEARCH USE

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

APPLICATIONS

NPC1 (C-21) is recommended for detection of Niemann-Pick type C1 protein (NPC1) of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

NPC1 (C-21) is also recommended for detection of Niemann-Pick type C1 protein (NPC1) in additional species, including equine, canine, bovine, porcine and avian.

Suitable for use as control antibody for NPC1 siRNA (h): sc-41588, NPC1 siRNA (m): sc-41589, NPC1 shRNA Plasmid (h): sc-41588-SH, NPC1 shRNA Plasmid (m): sc-41589-SH, NPC1 shRNA (h) Lentiviral Particles: sc-41588-V and NPC1 shRNA (m) Lentiviral Particles: sc-41589-V.

Molecular Weight of glycosylated NPC1: 170/190 kDa.

Positive Controls: ES-2 cell lysate: sc-24674.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

SELECT PRODUCT CITATIONS

1. Ong, W.Y., et al. 2004. Neuronal localization and association of Niemann Pick C2 protein (HE1/NPC2) with the postsynaptic density. Neuroscience 128: 561-570.
2. Dai, XY., et al. 2008. The effect of T0901317 on ATP-binding cassette transporter A1 and Niemann-Pick type C1 in ApoE$^{-/-}$ mice. J. Cardiovasc. Pharmacol. 51: 467-475.

STORAGE

Store at 4° C, **DO NOT FREEZE**. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

PROTOCOLS

See our web site at www.scbt.com or our catalog for detailed protocols and support products.