

NPC1L1 (S-18): sc-49065

BACKGROUND

Niemann-Pick disease type C (NPC) is an autosomal recessive disease characterized by the accumulation of unesterified cholesterol in the endosomal/lysosomal system, which results in progressive neurodegeneration and death. Niemann-Pick C1-like protein 1 precursor, or NPC1L1, is a membrane protein involved in the uptake of cholesterol at the intestinal enterocyte across the plasma membrane. NPC1L1 is widely expressed and is the target of ezetimibe, a drug involved in the inhibition of cholesterol absorption. In human, mouse and rat, small intestine tissue shows the highest level of NPC1L1 expression; expression in other tissues includes gallbladder, liver, testis and stomach. The NPC1L1 gene contains 20 exons, with an unusually large 1,526 bp exon 2, and spans approximately 29 kb. The presumed promoter region of the gene harbors a sterol-regulatory element (SRE) for SRE-binding protein, further suggesting that NPC1L1 may play a role in subcellular cholesterol homeostasis.

REFERENCES

- Davies, J.P., et al. 2005. Inactivation of NPC1L1 causes multiple lipid transport defects and protects against diet-induced hypercholesterolemia. *J. Biol. Chem.* 280: 12710-12720.
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- Hegele, R.A., et al. 2005. NPC1L1 haplotype is associated with inter-individual variation in plasma low-density lipoprotein response to ezetimibe. *Lipids Health Dis.* 4: 16.
- Iyer, S.P., et al. 2005. Characterization of the putative native and recombinant rat sterol transporter Niemann-Pick C1-like 1 (NPC1L1) protein. *Biochim. Biophys. Acta* 1722: 282-292.
- van der Veen, J.N., et al. 2005. Reduced cholesterol absorption upon PPAR α activation coincides with decreased intestinal expression of NPC1L1. *J. Lipid Res.* 46: 526-534.
- Wang, J., et al. 2005. Compound heterozygosity for two non-synonymous polymorphisms in NPC1L1 in a non-responder to ezetimibe. *Clin. Genet.* 67: 175-177.

CHROMOSOMAL LOCATION

Genetic locus: *Npc1l1* (mouse) mapping to 11 A1.

SOURCE

NPC1L1 (S-18) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the N-terminus of NPC1L1 of mouse origin.

PRODUCT

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

Blocking peptide available for competition studies, sc-49065 P, (100 μ g 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

NPC1L1 (S-18) is recommended for detection of NPC1L1 of mouse and rat 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).

Suitable for use as control antibody for NPC1L1 siRNA (m): sc-61226, NPC1L1 shRNA Plasmid (m): sc-61226-SH and NPC1L1 shRNA (m) Lentiviral Particles: sc-61226-V.

Molecular Weight of NPC1L1: 145 kDa.

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.

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.