SANTA CRUZ BIOTECHNOLOGY, INC.

NPC2 (V-16): sc-30347

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

Niemann-Pick disease, type C2 (NPC2), also known as epididymal secretory protein, is a secreted protein mapping against gene 14q24.3. NPC2 regulates the lipid composition of sperm membranes during maturation in the epididymis. Mutations in the NPC2 gene may cause Nieman-Pick type C2 disease and frontal lobe atrophy. Nieman-Pick type C2 is a fatal hereditary disease characterized by defective lysosome release of cholesterol. The disease is caused by HE1 deficiency, a lysosmal protein proven to be undetectable in fibroblasts from NPC2 patients. This differentiates NPC2 from NPC1, as NPC1 has HE1 protein present.

REFERENCES

1. Naureckiene, S. et al 2000. Identification of HE1 as the second gene of Niemann-Pick C disease. Science 290: 2298-2301.
2. Vanier, M.T. 2003. Niemann-Pick disease type C. Clin. Am. J. Hum. Genet. 64: 269-281.
3. Frolov, A. 2003. NPC1 and NPC2 regulate cellular cholesterol homeostasis through generation of low density lipoprotein cholesterol-derived oxysterols. J. Biol. Chem. 278: 25517-25525.
4. Ko, D.C. et al 2003. The integrity of a cholesterol-binding pocket in Niemann-Pick C2 protein is necessary to control lysosome cholesterol levels. Proc. Natl. Acad. Sci. USA 100: 2518-2525.
5. Sleat, D.E. 2004. Genetic evidence for nonredundant functional cooperativity between NPC1 and NPC2 in lipid transport. Proc. Natl. Acad. Sci. USA 101: 5886-5891.
6. Mutka, A.L. 2004. Secretion of sterols and the NPC2 protein from primary astrocytes. J. Biol. Chem. 279: 48654-48662.
7. Deisz, R.A. et al. 2005. Pathological cholesterol metabolism fails to modify electrophysiological properties of afflicted neurones in Niemann-Pick disease type C. Neuroscience 130: 867-873.

CHROMOSOMAL LOCATION

Genetic locus: NPC2 (human) mapping to 14q24.3.

SOURCE

NPC2 (V-16) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the C-terminus of NPC2 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-30347 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

STORAGE

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

APPLICATIONS

NPC2 (V-16) is recommended for detection of NPC2 of 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).

Suitable for use as control antibody for NPC2 siRNA (h): sc-43977, NPC2 shRNA Plasmid (h): sc-43977-SH and NPC2 shRNA (h) Lentiviral Particles: sc-43977-V.

Molecular Weight of NPC2: 16 kDa.

Positive Controls: NTERA-2 cl.D1 whole cell lysate: sc-364181, Caki-1 cell lysate: sc-2224 or Hep G2 cell lysate: sc-2227.

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.

RESEARCH USE

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

PROTOCOLS

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