

# NPC2 (I-14): sc-30349

## 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 lysosomal 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; Npc2 (mouse) mapping to 12 D1.

## SOURCE

NPC2 (I-14) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of NPC2 of mouse 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-30349 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 (I-14) is recommended for detection of NPC2 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).

Suitable for use as control antibody for NPC2 siRNA (h): sc-43977, NPC2 siRNA (m): sc-44816, NPC2 shRNA Plasmid (h): sc-43977-SH, NPC2 shRNA Plasmid (m): sc-44816-SH, NPC2 shRNA (h) Lentiviral Particles: sc-43977-V and NPC2 shRNA (m) Lentiviral Particles: sc-44816-V.

Molecular Weight of NPC2: 16 kDa.

Positive Controls: NTERA-2 cl.D1 cell lysate, Caki-1 cell lysate: sc-2224 or KNRK whole cell lysate: sc-2214.

## 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](http://www.scbt.com) or our catalog for detailed protocols and support products.



Try **NPC2 (D-3): sc-166449** or **NPC2 (H-10): sc-166321**, our highly recommended monoclonal alternatives to NPC2 (I-14).