

# NPC2 (H-10): sc-166321

## 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. 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.

## CHROMOSOMAL LOCATION

Genetic locus: NPC2 (human) mapping to 14q24.3; Npc2 (mouse) mapping to 12 D1.

## SOURCE

NPC2 (H-10) is a mouse monoclonal antibody raised against amino acids 21-145 mapping within an internal region of NPC2 of human origin.

## PRODUCT

Each vial contains 200 µg IgG<sub>2b</sub> kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

## APPLICATIONS

NPC2 (H-10) is recommended for detection of NPC2 of mouse, rat and human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500), immunohistochemistry (including paraffin-embedded sections) (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.

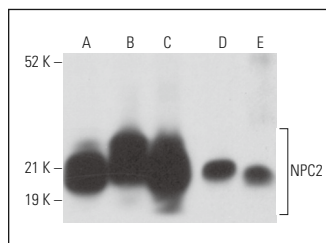
Molecular Weight of NPC2: 16 kDa.

Positive Controls: A-431 whole cell lysate: sc-2201, Hep G2 cell lysate: sc-2227 or SCC-4 whole cell lysate: sc-364363.

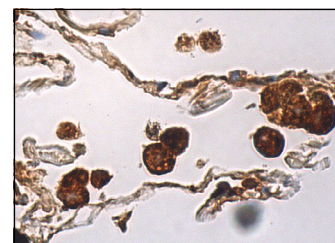
## RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850. 4) Immunohistochemistry: use m-IgGκ BP-HRP: sc-516102 with DAB, 50X: sc-24982 and Immunohistomount: sc-45086, or Organo/Limonene Mount: sc-45087.

## DATA



NPC2 (H-10): sc-166321. Western blot analysis of NPC2 expression in Hep G2 (A), A-431 (B), SCC-4 (C) and RPE-J (D) whole cell lysates and rat lung tissue extract (E).



NPC2 (H-10): sc-166321. Immunoperoxidase staining of formalin fixed, paraffin-embedded human lung tissue showing cytoplasmic staining of pneumocytes and macrophages.

## SELECT PRODUCT CITATIONS

1. Roszell, B.R., et al. 2012. Characterization of the Niemann-Pick C pathway in alveolar type II cells and lamellar bodies of the lung. *Am. J. Physiol. Lung Cell. Mol. Physiol.* 302: L919-L932.
2. Wu, C.C., et al. 2018. Integrated analysis of fine-needle-aspiration cystic fluid proteome, cancer cell secretome, and public transcriptome datasets for papillary thyroid cancer biomarker discovery. *Oncotarget* 9: 12079-12100.
3. Kubala, J.M., et al. 2023. NDUFA4L2 reduces mitochondrial respiration resulting in defective lysosomal trafficking in clear cell renal cell carcinoma. *Cancer Biol. Ther.* 24: 2170669.
4. Geng, F., et al. 2023. SREBP-1 upregulates lipophagy to maintain cholesterol homeostasis in brain tumor cells. *Cell Rep.* 42: 112790.

## STORAGE

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

## 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) for detailed protocols and support products.