NPC2 (H-125): sc-33776



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

- Naureckiene, S., et al. 2000. Identification of HE1 as the second gene of Niemann-Pick C disease. Science 290: 2298-2301.
- Vanier, M.T. 2003. Niemann-Pick disease type C. Clin. Am. J. Hum. Genet. 64: 269-281.
- 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.

CHROMOSOMAL LOCATION

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

SOURCE

NPC2 (H-125) is a rabbit polyclonal antibody raised against amino acids 21-145 mapping within an internal region of NPC2 of human origin.

PRODUCT

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

APPLICATIONS

NPC2 (H-125) 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), immunoprecipitation [1-2 μ g per 100-500 μ g of total protein (1 ml of cell lysate)], 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).

NPC2 (H-125) is also recommended for detection of NPC2 in additional species, including equine and porcine.

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.

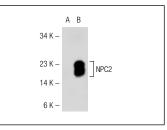
Molecular Weight of NPC2: 16 kDa.

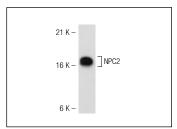
Positive Controls: NPC2 (m2): 293T Lysate: sc-122109, Caki-1 cell lysate: sc-2224 or NIH/3T3 whole cell lysate: sc-2210.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (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) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

DATA





NPC2 (H-125): sc-33776. Western blot analysis of NPC2 expression in non-transfected: sc-117752 (**A**) and mouse NPC2 transfected: sc-122109 (**B**) 293T whole rell lysates

NPC2 (H-125): sc-33776. Western blot analysis of NPC2 expression in NIH/3T3 whole cell lysate.

SELECT PRODUCT CITATIONS

- Araki, N., et al. 2009. Identification of NPC2 protein as interaction molecule with C2 domain of human Nedd4L. Biochem. Biophys. Res. Commun. 388: 290-296.
- Poirier, S., et al. 2013. The cytosolic adaptor AP-1A is essential for the trafficking and function of Niemann-Pick type C proteins. Traffic 14: 458-469.

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 or our catalog for detailed protocols and support products.

MONOS Satisfation Guaranteed

Try NPC2 (D-3): sc-166449 or NPC2 (H-10): sc-166321, our highly recommended monoclonal alternatives to NPC2 (H-125).