

NPC2 (D-3): sc-166449

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

- 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. 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.
- 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.
- Mutka, A.L. 2004. Secretion of sterols and the NPC2 protein from primary astrocytes. *J. Biol. Chem.* 279: 48654-48662.
- 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.

CHROMOSOMAL LOCATION

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

SOURCE

NPC2 (D-3) 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_{2a} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

NPC2 (D-3) is available conjugated to agarose (sc-166449 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-166449 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-166449 PE), fluorescein (sc-166449 FITC), Alexa Fluor® 488 (sc-166449 AF488), Alexa Fluor® 546 (sc-166449 AF546), Alexa Fluor® 594 (sc-166449 AF594) or Alexa Fluor® 647 (sc-166449 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-166449 AF680) or Alexa Fluor® 790 (sc-166449 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

RESEARCH USE

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

APPLICATIONS

NPC2 (D-3) 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) 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.

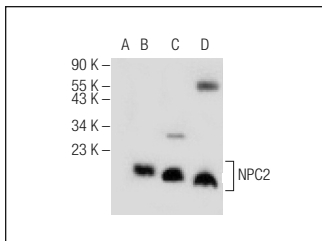
Molecular Weight of NPC2: 16 kDa.

Positive Controls: NIH/3T3 whole cell lysate: sc-2210, Hep G2 cell lysate: sc-2227 or NPC2 (m2): 293T Lysate: sc-122109.

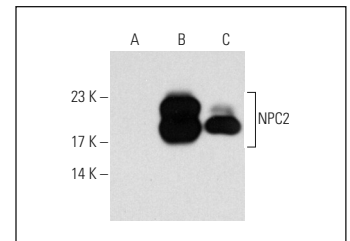
RECOMMENDED SUPPORT REAGENTS

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.

DATA



NPC2 (D-3): sc-166449. Western blot analysis of NPC2 expression in non-transfected CHO: sc-117750 (A), mouse NPC2 transfected CHO: sc-110157 (B), Hep G2 (C) and human PBL (D) whole cell lysates.



NPC2 (D-3): sc-166449. Western blot analysis of NPC2 expression in non-transfected 293T: sc-117752 (A), mouse NPC2 transfected 293T: sc-122109 (B) and NIH/3T3 (C) whole cell lysates.

SELECT PRODUCT CITATIONS

- Marquer, C., et al. 2016. Arf6 controls retromer traffic and intracellular cholesterol distribution via a phosphoinositide-based mechanism. *Nat. Commun.* 7: 11919.

STORAGE

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

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