

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

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.

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.

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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).

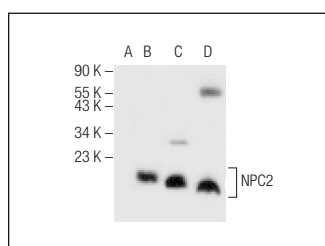
Suitable for use as control antibody for NPC2 siRNA (h): sc-43977, NPC2 siRNA (m): sc-44816, NPC2 shRNA Plasmid (h): sc-43977-SH, NPC2 shRNA Plasmid (m): sc-44816-SH, NPC2 shRNA (h) Lentiviral Particles: sc-43977-V and NPC2 shRNA (m) Lentiviral Particles: sc-44816-V.

Molecular Weight of NPC2: 16 kDa.

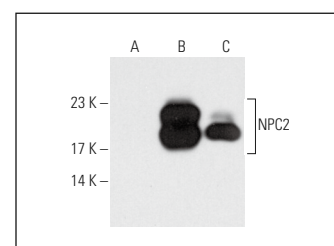
RECOMMENDED SUPPORT REAGENTS

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

1. Marquer, C., et al. 2016. Arf6 controls retromer traffic and intracellular cholesterol distribution via a phosphoinositide-based mechanism. *Nat. Commun.* 7: 11919.
2. Carna, M., et al. 2023. Pathogenesis of Alzheimer's disease: involvement of the choroid plexus. *Alzheimers Dement.* 19: 3537-3554.
3. Nara, A., et al. 2023. The ultrastructural function of MLN64 in the late endosome-mitochondria membrane contact sites in placental cells. *Exp. Cell Res.* 429: 113668.

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 for detailed protocols and support products.