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Produktinformation

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Lieferung & Zahlungsart

siehe unsere Liefer- und Versandbedingungen

Zuschläge

- Mindermengenzuschlag
- Trockeneiszuschlag
- Gefahrgutzuschlag
- Expressversand

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SANTA CRUZ BIOTECHNOLOGY, INC.

NPC2 (m2): 293T Lysate: sc-122109

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

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 (mouse) mapping to 12 D1.

PRODUCT

NPC2 (m2): 293T Lysate represents a lysate of mouse NPC2 transfected 293T cells and is provided as 100 µg protein in 200 µl SDS-PAGE buffer.

APPLICATIONS

NPC2 (m2): 293T Lysate is suitable as a Western Blotting positive control for mouse reactive NPC2 antibodies. Recommended use: 10-20 µl per lane.

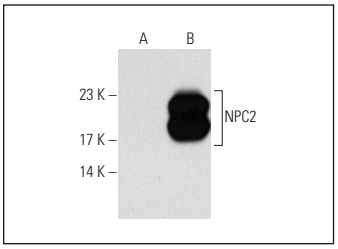
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

NPC2 (D-3): sc-166449 is recommended as a positive control antibody for Western Blot analysis of enhanced mouse NPC2 expression in NPC2 transfected 293T cells (starting dilution 1:100, dilution range 1:100-1:1,000).

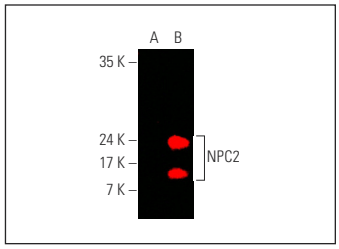
RECOMMENDED SUPPORT REAGENTS

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.

DATA



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



NPC2 (D-3) Alexa Fluor® 790: sc-166449 AF790. Direct near-infrared western blot analysis of NPC2 expression in non-transfected: sc-117752 (**A**) and mouse NPC2 transfected: sc-122109 (**B**) 293T whole cell lysates. Blocked with UltraCruz® Blocking Reagent: sc-516214.

STORAGE

Store at -20° C. Repeated freezing and thawing should be minimized. Sample vial should be boiled once prior to use. 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.