SANTA CRUZ BIOTECHNOLOGY, INC.

nephrocystin (D-9): sc-271190

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

Clinical features of familial juvenile nephronophthisis include anemia, polyuria, polydipsia, isosthenuria and death in uremia. Juvenile nephronophthisis type 1 is caused by mutations of NPHP1, the gene encoding for nephrocystin. Nephrocystin interacts with p130^{Cas} (BCAR1), proline-rich tyrosine kinase-2 (PTK2B or Pyk2) and tensin in embryonic kidney and testis, indicating that these proteins participate in a common signaling pathway. Nephrocystin and p130^{Cas} interact in mammalian cells and both proteins prominently localize at or near sites of cell-cell contact in polarized Madin-Darby canine kidney epithelial cells. Expression of nephrocystin results in phosphorylation of Pyk2 on Tyrosine 402 as well as activation of downstream mitogen-activated protein kinases, such as ERK1 and ERK2. Nephrocystin contains an SRC-homology 3 SH3 domain, which is highly conserved throughout evolution. The gene which encodes nephrocystin maps to human chromosome 2q13.

REFERENCES

1. Medhioub, M., Cherif, D., Benessy, F., Silbermann, F., Gubler, M.C., Le Paslier, D., Cohen, D., Weissenbach, J., Beckmann, J. and Antignac, C. 1994. Refined mapping of a gene (NPH1) causing familial juvenile nephronophthisis and evidence for genetic heterogeneity. Genomics 22: 296-301.
2. Donaldson, J.C., Dempsey, P.J., Reddy, S., Bouton, A.H., Coffey, R.J. and Hanks, S.K. 2000. Crk-associated substrate p130^{Cas} interacts with nephrocystin and both proteins localize to cell-cell contacts of polarized epithelial cells. Exp. Cell Res. 256: 168-178.
3. Benzing, T., Gerke, P., Hopker, K., Hildebrandt, F., Kim, E. and Walz, G. 2001. Nephrocystin interacts with Pyk2, p130^{Cas}, and tensin and triggers phosphorylation of Pyk2. Proc. Natl. Acad. Sci. USA 98: 9784-9789.
4. Hildebrandt, F. and Omram, H. 2001. New insights: nephronophthisis-medullary cystic kidney disease. Pediatr. Nephrol. 16: 168-176.

CHROMOSOMAL LOCATION

Genetic locus: NPHP1 (human) mapping to 2q13.

SOURCE

nephrocystin (D-9) is a mouse monoclonal antibody raised against amino acids 433-732 mapping at the C-terminus of nephrocystin of human origin.

PRODUCT

Each vial contains 200 µg IgG_1 kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

nephrocystin (D-9) is available conjugated to agarose (sc-271190 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-271190 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-271190 PE), fluorescein (sc-271190 FITC), Alexa Fluor® 488 (sc-271190 AF488), Alexa Fluor® 546 (sc-271190 AF546), Alexa Fluor® 594 (sc-271190 AF594) or Alexa Fluor® 647 (sc-271190 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-271190 AF680) or Alexa Fluor® 790 (sc-271190 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

APPLICATIONS

nephrocystin (D-9) is recommended for detection of nephrocystin isoforms 1-3 of 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 nephrocystin siRNA (h): sc-40769, nephrocystin shRNA Plasmid (h): sc-40769-SH and nephrocystin shRNA (h) Lentiviral Particles: sc-40769-V.

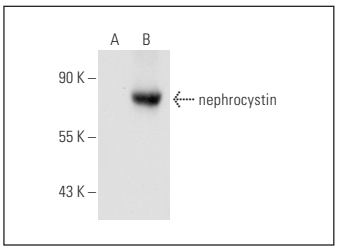
Molecular Weight of nephrocystin: 83 kDa.

Positive Controls: nephrocystin (h): 293T Lysate: sc-116755.

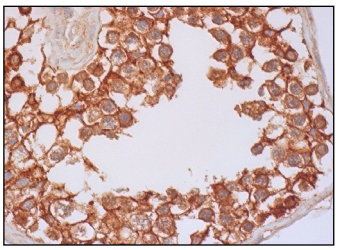
RECOMMENDED SUPPORT REAGENTS

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



nephrocystin (D-9): sc-271190. Western blot analysis of nephrocystin expression in non-transfected: sc-117752 (**A**) and human nephrocystin transfected: sc-116755 (**B**) 293T whole cell lysates.



nephrocystin (D-9): sc-271190. Immunoperoxidase staining of formalin fixed, paraffin-embedded human testis tissue showing membrane and cytoplasmic staining of cells in seminiferous ducts.

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.