

# nephrocystin (D-9): sc-271190

## 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<sup>Cas</sup> (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<sup>Cas</sup> 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 (NPHP1) 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<sup>Cas</sup> 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<sup>Cas</sup>, 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<sub>1</sub> 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.

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## 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.

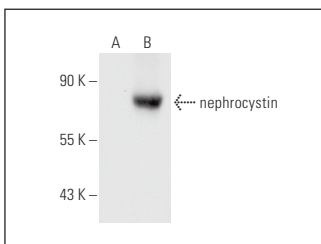
Molecular Weight of nephrocystin: 83 kDa.

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

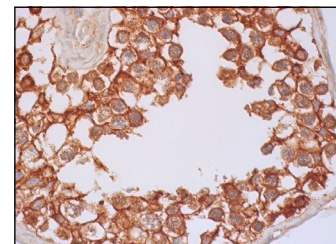
## RECOMMENDED SUPPORT REAGENTS

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.