

# nephrocystin (C-20): sc-20204

## 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 a 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., et al. 1994. Refined mapping of a gene (NPH1) causing familial juvenile nephronophthisis and evidence for genetic heterogeneity. *Genomics* 22: 296-301.
2. Donaldson, J.C., et al. 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., et al. 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., et al. 2001. New insights: nephronophthisis-medullary cystic kidney disease. *Pediatr. Nephrol.* 16: 168-176.
5. LocusLink Report (LocusID: 256100). <http://www.ncbi.nlm.nih.gov/LocusLink>

## CHROMOSOMAL LOCATION

Genetic locus: NPHP1 (human) mapping to 2q13; Nphp1 (mouse) mapping to 2 F1.

## SOURCE

nephrocystin (C-20) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the C-terminus of nephrocystin of human origin.

## PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-20204 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

## STORAGE

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

## APPLICATIONS

nephrocystin (C-20) is recommended for detection of nephrocystin of mouse, rat and human origin by Western Blotting (starting dilution 1:200, 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).

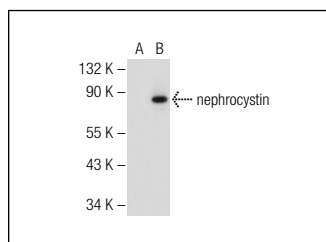
nephrocystin (C-20) is also recommended for detection of nephrocystin in additional species, including canine and porcine.

Suitable for use as control antibody for nephrocystin siRNA (h): sc-40769, nephrocystin siRNA (m): sc-40770, nephrocystin shRNA Plasmid (h): sc-40769-SH, nephrocystin shRNA Plasmid (m): sc-40770-SH, nephrocystin shRNA (h) Lentiviral Particles: sc-40769-V and nephrocystin shRNA (m) Lentiviral Particles: sc-40770-V.

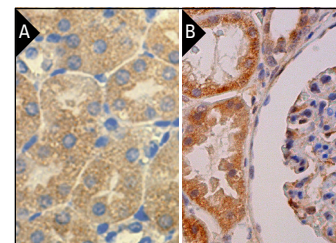
Molecular Weight of nephrocystin: 83 kDa.

Positive Controls: mouse kidney extract: sc-2255, nephrocystin (h): 293T Lysate: sc-116755 or mouse embryo extract: sc-364239.

## DATA



nephrocystin (C-20): sc-20204. Western blot analysis of nephrocystin expression in non-transfected: sc-117752 (A) and human nephrocystin transfected: sc-116755 (B) 293T whole cell lysates.



nephrocystin (C-20): sc-20204. Immunoperoxidase staining of formalin-fixed, paraffin-embedded mouse kidney tissue showing cell junction localization (A). Immunoperoxidase staining of formalin fixed, paraffin-embedded human kidney tissue showing cytoplasmic staining of cells in glomeruli and cells in tubules (B).

## SELECT PRODUCT CITATIONS

1. Eley, L., et al. 2008. Nephrocystin-1 interacts directly with Ack1 and is expressed in human collecting duct. *Biochem. Biophys. Res. Commun.* 371: 877-882.

## RESEARCH USE

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

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Try **nephrocystin (D-9): sc-271190**, our highly recommended monoclonal alternative to nephrocystin (C-20).