

# nephrocystin (H-300): sc-33592

## 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 p130Cas (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 p130Cas 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.

## CHROMOSOMAL LOCATION

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

## SOURCE

nephrocystin (H-300) is a rabbit polyclonal antibody raised against amino acids 433-732 mapping at 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.

## STORAGE

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

## APPLICATIONS

nephrocystin (H-300) is recommended for detection of nephrocystin isoforms 1-3 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) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

nephrocystin (H-300) is also recommended for detection of nephrocystin isoforms 1-3 in additional species, including equine, canine, bovine 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.

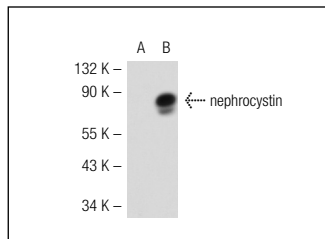
Molecular Weight of nephrocystin: 83 kDa.

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

## RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 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 goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

## DATA



nephrocystin (H-300): sc-33592. Western blot analysis of nephrocystin expression in non-transfected: sc-117752 (A) and human nephrocystin transfected: sc-116557 (B) 293T whole cell lysates.

## SELECT PRODUCT CITATIONS

- Eley, L., et al. 2008. Joubertin localizes to collecting ducts and interacts with nephrocystin-1. *Kidney Int.* 9: 1139-1149.

## RESEARCH USE

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

## PROTOCOLS

See our web site at [www.scbt.com](http://www.scbt.com) or our catalog for detailed protocols and support products.



Try **nephrocystin (D-9): sc-271190**, our highly recommended monoclonal alternative to nephrocystin (H-300).