

# nephrocystin-5 (E-18): sc-51342

## BACKGROUND

The nephrocystin proteins comprise a family of five enzymes that commonly interact with p130 Cas, proline-rich tyrosine kinases, calmodulin and tensin, indicating that these proteins may participate in a common signaling pathway. Localized to the outer segments and primary cilia of photoreceptor cells, nephrocystin-5 is complexed with RPGR (retinitis pigmentosa GTPase regulator) and interacts directly with calmodulin. Nephrocystin-5 is thought to participate with RPGR in a pathway of ciliary function in the kidney and retina. Mutations in the gene encoding nephrocystin-5 are the primary cause of Senior-Loken syndrome 5, a juvenile disorder characterized by defects in the waste filtering system of the kidney, as well as retinal degradation.

## REFERENCES

1. Online Mendelian Inheritance in Man, OMIM™. 2002. Johns Hopkins University, Baltimore, MD. MIM Number: 609237. World Wide Web URL: <http://www.ncbi.nlm.nih.gov/omim/>
2. Otto, E.A., Loeys, B., Khanna, H., Hellemans, J., Sudbrak, R., Fan, S., Muerb, U., O'Toole, J.F., Helou, J., Attanasio, M., Utsch, B., Sayer, J.A., Lillo, C., Jimeno, D., Coucke, P., De Paepe, A., Reinhardt, R., Klages, S., Tsuda, M., Kawakami, I., Kusakabe, T., Omran, H., et al. 2005. Nephrocystin-5, a ciliary IQ domain protein, is mutated in Senior-Loken syndrome and interacts with RPGR and calmodulin. *Nat. Genet.* 37: 282-288.
3. Fliegau, M., Horvath, J., von Schnakenburg, C., Olbrich, H., Müller, D., Thumfart, J., Schermer, B., Pazour, G.J., Neumann, H.P., Zentgraf, H., Benzing, T. and Omran, H. 2006. Nephrocystin specifically localizes to the transition zone of renal and respiratory cilia and photoreceptor connecting cilia. *J. Am. Soc. Nephrol.* 17: 2424-2433.
4. Guyon, R., Pearce-Kelling, S.E., Zeiss, C.J., Acland, G.M. and Aguirre, G.D. 2007. Analysis of six candidate genes as potential modifiers of disease expression in canine XLPRA1, a model for human X-linked retinitis pigmentosa 3. *Mol. Vis.* 13: 1094-1105.
5. von Schnakenburg, C., Fliegau, M. and Omran, H. 2007. Nephrocystin and ciliary defects not only in the kidney? *Pediatr. Nephrol.* 22: 765-769.

## CHROMOSOMAL LOCATION

Genetic locus: IQCB1 (human) mapping to 3q13.33; *lqcb1* (mouse) mapping to 16 B3.

## SOURCE

nephrocystin-5 (E-18) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the N-terminus of nephrocystin-5 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-51342 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

## APPLICATIONS

nephrocystin-5 (E-18) is recommended for detection of nephrocystin-5 isoforms 1, 2 and 3 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), 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-5 (E-18) is also recommended for detection of nephrocystin-5 isoforms 1, 2 and 3 in additional species, including equine, canine, bovine and porcine.

Suitable for use as control antibody for nephrocystin-5 siRNA (h): sc-72270, nephrocystin-5 siRNA (m): sc-72271, nephrocystin-5 shRNA Plasmid (h): sc-72270-SH, nephrocystin-5 shRNA Plasmid (m): sc-72271-SH, nephrocystin-5 shRNA (h) Lentiviral Particles: sc-72270-V and nephrocystin-5 shRNA (m) Lentiviral Particles: sc-72271-V.

Molecular Weight of nephrocystin-5: 69 kDa.

## RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (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) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

## 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](http://www.scbt.com) or our catalog for detailed protocols and support products.