

nephrocystin-4 (C-13): sc-49244

BACKGROUND

The nephrocystin proteins comprise a family of five enzymes that commonly interact with p130Cas, proline-rich tyrosine kinases, calmodulin, and tensin, indicating that these proteins may participate in a common signaling pathway. nephrocystin-4 is a 1,250-amino acid protein that interacts with signaling molecules involved in cell adhesion and organization of the actin cytoskeleton, such as Pyk2, tensin, and filamins. nephrocystin-4 colocalizes with PKD-2 in the transition zones of ciliated sensory endings of dendrites, and, together, they play an important role in facilitating ciliary sensory signal transduction. Mutations in the nephrocystin-4 gene contribute to the disease nephronophthisis, an autosomal-recessive cystic kidney disease. Clinical features of familial juvenile nephronophthisis include anemia, polyuria, polydipsia, isosthenuria, and death.

REFERENCES

- Mollet, G., et al. 2002. The gene mutated in juvenile nephronophthisis type 4 encodes a novel protein that interacts with nephrocystin. *Nat. Genet.* 32: 300-305.
- Online Mendelian Inheritance in Man, OMIM™. 2002. Johns Hopkins University, Baltimore, MD. MIM Number: 607215. World Wide Web URL: <http://www.ncbi.nlm.nih.gov/omim/>
- Olbrich, H., et al. 2003. Mutations in a novel gene, NPHP3, cause adolescent nephronophthisis, tapeto-retinal degeneration and hepatic fibrosis. *Nat. Genet.* 34: 455-459.
- Hoefele, J., et al. 2004. Clinical and histological presentation of three siblings with mutations in the NPHP4 gene. *Am. J. Kidney Dis.* 43: 358-364.
- Jauregui, A.R., et al. 2005. Functional characterization of the *C. elegans* nephrocystins NPHP-1 and NPHP-4 and their role in cilia and male sensory behaviors. *Exp. Cell Res.* 305: 333-342.
- Mollet, G., et al. 2005. Characterization of the nephrocystin/nephrocystin-4 complex and subcellular localization of Nephrocystin-4 to primary cilia and centrosomes. *Hum. Mol. Genet.* 14: 645-656.
- Roepman, R., et al. 2005. Interaction of nephrocystin-4 and RPGRIP1 is disrupted by nephronophthisis or Leber congenital amaurosis-associated mutations. *Proc. Natl. Acad. Sci. USA* 102: 18520-18525.
- Winkelbauer, M.E., et al. 2005. The *C. elegans* homologs of nephrocystin-1 and nephrocystin-4 are cilia transition zone proteins involved in chemosensory perception. *J. Cell Sci.* 118: 5575-5587.
- Wolf, M.T., et al. 2005. Expression and phenotype analysis of the nephrocystin-1 and nephrocystin-4 homologs in *Caenorhabditis elegans*. *J. Am. Soc. Nephrol.* 16: 676-687.

CHROMOSOMAL LOCATION

Genetic locus: NPHP4 (human) mapping to 1p36.31; Nphp4 (mouse) mapping to 4 E2.

STORAGE

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

SOURCE

nephrocystin-4 (C-13) is an affinity purified goat polyclonal antibody raised against a peptide mapping at the C-terminus of nephrocystin-4 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-49244 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

nephrocystin-4 (C-13) is recommended for detection of nephrocystin-4 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-4 (C-13) is also recommended for detection of nephrocystin-4 in additional species, including equine, canine and bovine.

Suitable for use as control antibody for nephrocystin-4 siRNA (h): sc-61182, nephrocystin-4 siRNA (m): sc-61183, nephrocystin-4 shRNA Plasmid (h): sc-61182-SH, nephrocystin-4 shRNA Plasmid (m): sc-61183-SH, nephrocystin-4 shRNA (h) Lentiviral Particles: sc-61182-V and nephrocystin-4 shRNA (m) Lentiviral Particles: sc-61183-V.

Molecular Weight of nephrocystin-4: 175 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.

RESEARCH USE

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

PROTOCOLS

See our web site at www.scbt.com or our catalog for detailed protocols and support products.