

nephrocystin-4 (H-67): sc-99012

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. 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

1. 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.
2. 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/>
3. 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.
4. Hoefele, J., et al. 2004. Clinical and histological presentation of 3 siblings with mutations in the NPHP4 gene. *Am. J. Kidney Dis.* 43: 358-364.
5. 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.
6. 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.

CHROMOSOMAL LOCATION

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

SOURCE

nephrocystin-4 (H-67) is a rabbit polyclonal antibody raised against amino acids 1116-1182 mapping within an internal region 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.

STORAGE

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

APPLICATIONS

nephrocystin-4 (H-67) 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), 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-4 (H-67) 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.

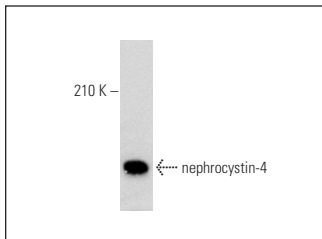
Molecular Weight of nephrocystin-4: 175 kDa.

Positive Controls: mouse brain extract: sc-2253.

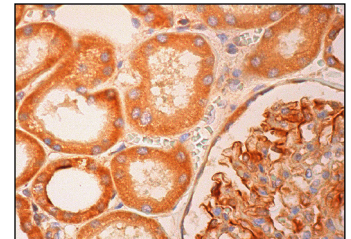
RECOMMENDED SECONDARY REAGENTS

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. 4) Immunohistochemistry: use ImmunoCruz™: sc-2051 or ABC: sc-2018 rabbit IgG Staining Systems.

DATA



nephrocystin-4 (H-67): sc-99012. Western blot analysis of nephrocystin-4 expression in mouse brain tissue extract.



nephrocystin-4 (H-67): sc-99012. Immunoperoxidase staining of formalin fixed, paraffin-embedded human kidney tissue showing membrane and cytoplasmic staining of cells in glomeruli and cytoplasmic staining of cells in tubules.

RESEARCH USE

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