

nephrocystin-3 (C-19): sc-49458

BACKGROUND

The nephrocystin proteins comprise a family of five enzymes that commonly interact with p130Cas (BCAR1), proline-rich tyrosine kinases, calmodulin and tensin, indicating that these proteins may participate in a common signaling pathway. The NPHP3 gene encodes nephrocystin-3, a protein that interacts with nephrocystin and may play a role in renal tubular function and development. nephrocystin-3 contains a tubulin-tyrosine ligase (TTL) domain, a coiled-coil (CC) domain and a tetratricopeptide repeat (TPR) domain. It is expressed widely at a low level, specifically in tissues such as the heart, placenta, liver, skeletal muscle, kidney and pancreas. In the brain and lung, nephrocystin-3 is expressed at a very low level. Mutations in NPHP3 may cause nephronophthisis type 3, a recessive disorder affecting adolescents characterized by sclerosing tubulointerstitial nephropathy, alterations of tubular basement membranes, tubular atrophy and dilatation, and renal cyst development primarily at the corticomedullary junction. These symptoms lead to chronic renal failure in affected individuals.

REFERENCES

1. Omran, H., et al. 2000. Identification of a new gene locus for adolescent nephronophthisis, on chromosome 3q22 in a large Venezuelan pedigree. *Am. J. Hum. Genet.* 66: 118-127.
2. Omran, H., et al. 2001. Human adolescent nephronophthisis: gene locus synteny disease in pcy mice. *J. Am. Soc. Nephrol.* 12: 107-113.
3. Online Mendelian Inheritance in Man, OMIM[™]. 2002. Johns Hopkins University, Baltimore, MD. MIM Number: 608002. World Wide Web URL: <http://www.ncbi.nlm.nih.gov/omim/>
4. Olbrich, H., et al. 2003. Mutat degeneration and hepatic fibrosis. *Nat. Genet.* 34: 455-459.
5. Tanner, J.A., et al. 2004. Dietary potassium citrate does not harm the pcy mouse. *Exp. Biol. Med.* 230: 57-60.

CHROMOSOMAL LOCATION

Genetic locus: NPHP3 (human) mapping to 3q22.1; Nphp3 (mouse) mapping to 9 F1.

SOURCE

nephrocystin-3 (C-19) is an affinity purified goat polyclonal antibody raised against a peptide mapping at the C-terminus of nephrocystin-3 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-49458 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-3 (C-19) is recommended for detection of nephrocystin-3 isoforms 1 and 2 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-3 (C-19) is also recommended for detection of nephrocystin-3 isoforms 1 and 2 in additional species, including canine.

Suitable for use as control antibody for nephrocystin-3 siRNA (h): sc-61180, nephrocystin-3 siRNA (m): sc-61181, nephrocystin-3 shRNA Plasmid (h): sc-61180-SH, nephrocystin-3 shRNA Plasmid (m): sc-61181-SH, nephrocystin-3 shRNA (h) Lentiviral Particles: sc-61180-V and nephrocystin-3 shRNA (m) Lentiviral Particles: sc-61181-V.

Molecular Weight of nephrocystin-3: 148 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.