

nephrocystin-3 (3B1): sc-517129

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. and Tanner, G.A. 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 (3B1) is a mouse monoclonal antibody raised against amino acids 106-205 representing partial length nephrocystin-3 of human origin.

PRODUCT

Each vial contains 100 µg IgG₁ kappa light chain in 1.0 ml 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.

RESEARCH USE

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

APPLICATIONS

nephrocystin-3 (3B1) is recommended for detection of nephrocystin-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)] and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

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.

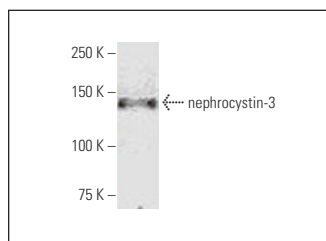
Positive Controls: Hep G2 cell lysate: sc-2227.

RECOMMENDED SUPPORT REAGENTS

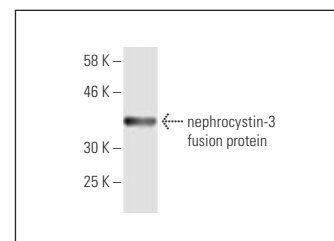
To ensure optimal results, the following support reagents are recommended:

- 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), 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).

DATA



nephrocystin-3 (3B1); sc-517129. Western blot analysis of nephrocystin-3 expression in Hep G2 whole cell lysate.



nephrocystin-3 (3B1); sc-517129. Western blot analysis of human recombinant nephrocystin-3 fusion protein.

PROTOCOLS

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