

nephrocystin (T-20): sc-20206

BACKGROUND

Clinical features of familial juvenile nephronophthisis include anemia, polyuria, polydipsia, isosthenuria, and death in uremia. Juvenile nephronophthisis type 1 is caused by mutations of NPHP1, the gene encoding for nephrocystin. Nephrocystin interacts with p130Cas (BCAR1), proline-rich tyrosine kinase-2 (PTK2B or Pyk2), and tensin in embryonic kidney and testis, indicating that these proteins participate in a common signaling pathway. Nephrocystin and p130Cas interact in mammalian cells and both proteins prominently localize at or near sites of cell-cell contact in polarized Madin-Darby canine kidney epithelial cells. Expression of nephrocystin results in phosphorylation of Pyk2 on Tyrosine 402 as well as activation of downstream mitogen-activated protein kinases, such as ERK1 and ERK2. Nephrocystin contains a src-homology 3 (SH₃) domain, which is highly conserved throughout evolution. The gene which encodes nephrocystin maps to human chromosome 2q13.

REFERENCES

1. LocusLink Report (LocusID: 256100). <http://www.ncbi.nlm.nih.gov/LocusLink>
2. Medhioub, M., et al. 1994. Refined mapping of a gene (NPH1) causing familial juvenile nephronophthisis and evidence for genetic heterogeneity. *Genomics* 22: 296-301.
3. Donaldson, J.C., et al. 2000. Crk-associated substrate p130(Cas) interacts with nephrocystin and both proteins localize to cell-cell contacts of polarized epithelial cells. *Exp. Cell Res.* 256: 168-178.
4. Benzing, T., et al. 2001. Nephrocystin interacts with Pyk2, p130(Cas), and tensin and triggers phosphorylation of Pyk2. *Proc. Natl. Acad. Sci. USA* 98: 9784-9789.
5. Hildebrandt, F., et al. 2001. New insights: nephronophthisis-medullary cystic kidney disease. *Pediatr. Nephrol.* 16: 168-176.

SOURCE

nephrocystin (T-20) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of nephrocystin of mouse 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-20206 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.

PROTOCOLS

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

APPLICATIONS

nephrocystin (T-20) is recommended for detection of nephrocystin of mouse and rat 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) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for nephrocystin siRNA (m): sc-40770.

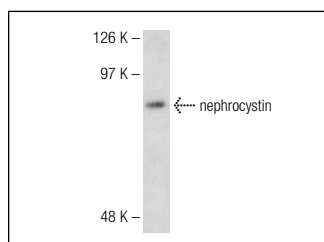
Molecular Weight of nephrocystin: 83 kDa.

Positive Controls: mouse kidney extract: sc-2255.

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) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/ 2.0 ml). 3) 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.

DATA



nephrocystin (T-20): sc-20206. Western blot analysis of nephrocystin expression in mouse kidney tissue extract.

RESEARCH USE

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