SANTA CRUZ BIOTECHNOLOGY, INC.

nephrocystin (h): 293T Lysate: sc-116755

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 an SRC-homology 3 SH3 domain, which is highly conserved throughout evolution. The gene which encodes nephrocystin maps to human chromosome 2q13.

REFERENCES

1. Medhioub, M., Cherif, D., Benessy, F., Silbermann, F., Gubler, M.C., Le Paslier, D., Cohen, D., Weissenbach, J., Beckmann, J. and Antignac, C. 1994. Refined mapping of a gene (NPH1) causing familial juvenile nephronophthisis and evidence for genetic heterogeneity. Genomics 22: 296-301.
2. Donaldson, J.C., Dempsey, P.J., Reddy, S., Bouton, A.H., Coffey, R.J. and Hanks, S.K. 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.
3. Benzing, T., Gerke, P., Hopker, K., Hildebrandt, F., Kim, E. and Walz, G. 2001. Nephrocystin interacts with Pyk2, p130^{Cas}, and tensin and triggers phosphorylation of Pyk2. Proc. Natl. Acad. Sci. USA 98: 9784-9789.
4. Hildebrandt, F. and Omram, H. 2001. New insights: nephronophthisis-medullary cystic kidney disease. Pediatr. Nephrol. 16: 168-176.
5. LocusLink Report (LocusID: 256100). http://www.ncbi.nlm.nih.gov/LocusLink/

CHROMOSOMAL LOCATION

Genetic locus: NPHP1 (human) mapping to 2q13.

PRODUCT

nephrocystin (h): 293T Lysate represents a lysate of human nephrocystin transfected 293T cells and is provided as 100 µg protein in 200 µl SDS-PAGE buffer.

APPLICATIONS

nephrocystin (h): 293T Lysate is suitable as a Western Blotting positive control for human reactive nephrocystin antibodies. Recommended use: 10-20 µl per lane.

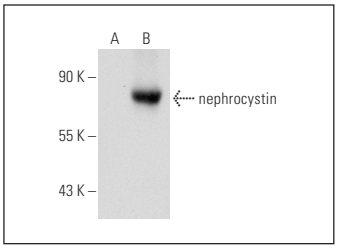
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

nephrocystin (D-9): sc-271190 is recommended as a positive control antibody for Western Blot analysis of enhanced human nephrocystin expression in nephrocystin transfected 293T cells (starting dilution 1:100, dilution range 1:100-1:1,000).

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, UltraCruz® Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048.

DATA



nephrocystin (D-9): sc-271190. Western blot analysis of nephrocystin expression in non-transfected: sc-117752 (**A**) and human nephrocystin transfected: sc-116755 (**B**) 293T whole cell lysates.

STORAGE

Store at -20° C. Repeated freezing and thawing should be minimized. Sample vial should be boiled once prior to use. Non-hazardous. No MSDS required.

RESEARCH USE

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

PROTOCOLS

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