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SZABO-SCANDIC HandelsgmbH

Quellenstraße 110, A-1100 Wien

T. +43(0)1 489 3961-0

F. +43(0)1 489 3961-7

mail@szabo-scandic.com

www.szabo-scandic.com

[linkedin.com/company/szaboscandic](https://www.linkedin.com/company/szaboscandic) 

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

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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.
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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.

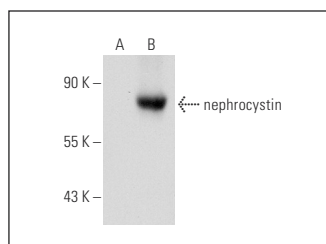
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

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.