



Product Information

Anti-Nephronophthisis (NPH) (pic)

Developed in Rabbit, IgG Fraction of Antiserum

Product Number **N6523**

Product Description

Anti-Nephronophthisis (NPH, NPHP1, nephrocystin) (pic) is developed in rabbit using a highly purified fusion protein of GST and a portion of mouse NPH corresponding to amino acid residues 612-691 (pic region) as the immunogen.

Anti-Nephronophthisis (pic) recognizes the pic region of mouse NPH protein (90 kDa) by immunoblotting and immunoprecipitation.

Juvenile nephronophthisis, an autosomal recessive cystic kidney disease, is the primary cause for chronic renal failure in children. The gene for nephronophthisis type 1 (NPHP1) has recently been identified.¹ Its gene product, nephrocystin, is a novel protein of unknown function, which contains a src-homology 3 domain.

NPHP1 expression has been studied in mouse and human systems.² Expression in the human adult is widespread but relatively weak. In contrast, expression in adult mouse is strong and localized to the testis. *In situ* hybridization to whole mouse embryo demonstrates widespread and consistently strong expression throughout all developmental stages.

It is hypothesized that the NPHP1 gene product, nephrocystin, is involved in signaling processes at focal adhesions (cell and extracellular matrix contact) and/or adherens junctions (cell-cell contact).³ The hypothesis is based on the fact that most src-homology 3 -containing proteins are part of focal adhesion signaling complexes. Additionally, nephrocystin binds to p130(cas), a prominent participant in focal adhesion signaling.

Reagents

Anti-Nephronophthisis (pic) is supplied as 250 µg of purified IgG at a concentration of 0.2 µM in phosphate buffered saline with 0.08% sodium azide.

Precautions and Disclaimer

Due to the sodium azide content, a material safety data sheet (MSDS) for this product has been sent to the attention of the safety officer of your institution. Consult the MSDS for information regarding hazardous and safe handling practices.

Storage/Stability

Antibodies should be stored at -20 °C. For extended storage, freeze in working aliquots. Repeated freezing and thawing is not recommended. Storage in "frost-free" freezers is not recommended. If slight turbidity occurs upon prolonged storage, clarify the solution by centrifugation before use. Working dilution samples should be discarded if not used within 12 hours.

Product Profile

The recommended working dilution is 2-10 µg/ml for immunoblotting using peroxidase conjugated goat anti-rabbit IgG and chemiluminescent detection.

The recommended working dilution is 2-10 µg/ml for immunoprecipitation.

Note: In order to obtain best results and assay sensitivities of various techniques and preparations, we recommend determining optimal working dilutions by titration.

References

1. Hildebrandt, F. et al., Nat. Genet., **17**, 149-153 (1997).
2. Otto, E. et al., J. Am. Soc. Nephrol., **11**, 270-282 (2000).
3. Hildebrandt, F. and Otto, E., J. Am. Soc. Nephrol., **11**, 1753-1761 (2000).

kaa/mje 11/03

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