

Product datasheet for **TA335137**

Nephronophthisis (NPHP1) Rabbit Polyclonal Antibody

Product data:

Product Type:	Primary Antibodies
Applications:	WB
Recommended Dilution:	WB
Reactivity:	Human
Host:	Rabbit
Isotype:	IgG
Clonality:	Polyclonal
Immunogen:	The immunogen for anti-NPHP1 antibody: synthetic peptide directed towards the middle region of human NPHP1. Synthetic peptide located within the following region: GILFELGISYIRNSTGERGELSCGWVFLKLFDA SGVPIPAKTYELFLNGG
Formulation:	Liquid. Purified antibody supplied in 1x PBS buffer with 0.09% (w/v) sodium azide and 2% sucrose. <i>Note that this product is shipped as lyophilized powder to China customers.</i>
Purification:	Affinity Purified
Conjugation:	Unconjugated
Storage:	Store at -20°C as received.
Stability:	Stable for 12 months from date of receipt.
Predicted Protein Size:	83 kDa
Gene Name:	nephrocystin 1
Database Link:	NP_000263 Entrez Gene 4867 Human O15259



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Background:

Together with Cas NPHP1 may play a role in the control of epithelial cell polarity. NPHP1 seems to help to recruit protein tyrosine kinase 2 beta (PTK2B) to cell matrix adhesions, thereby initiating phosphorylation of PTK2B and PTK2B-dependent signaling. This gene encodes a protein with src homology domain 3 (SH3) patterns. This protein interacts with Crk-associated substrate, and it appears to function in the control of cell division, as well as in cell-cell and cell-matrix adhesion signaling, likely as part of a multifunctional complex localized in actin- and microtubule-based structures. Mutations in this gene cause familial juvenile nephronophthisis type 1, a kidney disorder involving both tubules and glomeruli. Defects in this gene are also associated with Senior-Loken syndrome type 1, also referred to as juvenile nephronophthisis with Leber amaurosis, which is characterized by kidney and eye disease, and with Joubert syndrome type 4, which is characterized by cerebellar ataxia, oculomotor apraxia, psychomotor delay and neonatal breathing abnormalities, sometimes including retinal dystrophy and renal disease. Multiple transcript variants encoding different isoforms have been found for this gene.

Synonyms:

JBTS4; NPH1; SLSN1

Note:

Immunogen Sequence Homology: Dog: 100%; Pig: 100%; Rat: 100%; Horse: 100%; Human: 100%; Mouse: 100%; Bovine: 100%; Rabbit: 100%; Guinea pig: 93%; Zebrafish: 92%

Protein Families:

Druggable Genome

Product images:


WB Suggested Anti-NPHP1 Antibody Titration:
0.2-1 ug/ml; ELISA Titer: 1:1562500; Positive
Control: Human heart