

Rabbit Anti-Inversin antibody

SL12437R

Product Name:	Inversin
Chinese Name:	内脏器官发育转位相关蛋白NPH2抗体
Alias:	INV; Inversion of embryo turning homolog; inversion of embryonic turning; INVS; Nephrocystin 2; Nephrocystin-2; Nephrocystin2; nephronophthisis 2 (infantile); NPH2; NPHP2; INVS_HUMAN.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human, Mouse, Rat, Cow, Sheep, Chimpanzee,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100- 500IF=1:100-500 (Paraffin sections need antigen repair) not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	118kDa 🧹
Cellular localization:	cytoplasmicThe cell membrane
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human Inversin/Nephrocystin 2:31- 130/1065
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
PubMed:	PubMed
Product Detail:	Nephrocystin-2 is a 1,065 amino acid protein that exists as three alternatively spliced isoforms and is essential for establishment of the left-right axis and normal renal development. Localizing to the cytoplasm, cytoskeleton, membrane and nucleus, nephrocystin-2 is expressed during presomite-stage embryos and persists in adulthood,

with high levels of expression in liver and kidney. Mice expressing nephrocystin-2 mutations are primarily generated by random insertional mutagenesis and result in the reversal of left/right polarity and cyst formation in the kidneys. Furthermore, altered nephrocystin-2 function reverses nodal and lefty expression, indicating that nephrocystin-2 signaling occurs upstream of these proteins involved in the development of asymmetry.

Function:

Required for normal renal development and establishment of left-right axis. Probably acts as a molecular switch between different Wnt signaling pathways. Inhibits the canonical Wnt pathway by targeting cytoplasmic disheveled (DVL1) for degradation by the ubiquitin-proteasome. This suggests that it is required in renal development to oppose the repression of terminal differentiation of tubular epithelial cells by Wnt signaling. Binds calmodulin via its IQ domains. Interacts with microtubules. (from SwissProt).

Subunit:

Binds calmodulin via its IQ domains. Interacts with APC2. Interacts with alpha-, beta-, and gamma-catenin. Interacts with N-cadherin (CDH2). Interacts with microtubules (By similarity). Interacts with NPHP1. Interacts with DVL1, PRICKLE (PRICKLE1 or PRICKLE2) and Strabismus (VANGL1 or VANGL2). Interacts with NPHP3. Interacts with IQCB1; the interaction likely requires additional interactors.

Subcellular Location:

Cytoplasm, cytoskeleton, spindle, membrane; Peripheral membrane protein, nucleus. Note=Associates with several components of the cytoskeleton including ciliary, random and polarized microtubules. During mitosis, it is recruited to mitotic spindle. Frequently membrane-associated, membrane localization is dependent upon cell-cell contacts and is redistributed when cell adhesion is disrupted after incubation of the cell monolayer with low-calcium/EGTA medium.

Tissue Specificity:

Widely expressed. Strongly expressed in the primary cilia of renal tubular cells.

Post-translational modifications:

May be ubiquitinated via its interaction with APC2 (By similarity).

DISEASE:

Defects in INVS are the cause of nephronophthisis type 2 (NPHP2) [MIM:602088]; also known as infantile nephronophthisis. NPHP2 is an autosomal recessive disorder resulting in end-stage renal disease. It is characterized by early onset and rapid progression. Phenotypic manifestations include enlarged kidneys, chronic tubulo-interstitial nephritis, anemia, hyperkalemic metabolic acidosis. Some patients also display situs inversus. Pathologically, it differs from later-onset nephronophthisis by the absence of medullary cysts and thickened tubular basement membranes and by the presence of cortical microcysts.

Similarity:

Contains 16 ANK repeats. Contains 2 IQ domains.

SWISS: Q9Y283

Gene ID: 27130

Database links:

Entrez Gene: 27130 Human

Entrez Gene: 16348 Mouse

Entrez Gene: 313228 Rat

<u>Omim: 243305</u> Human

SwissProt: Q9Y283 Human

SwissProt: O89019 Mouse

Unigene: 558477 Human

Important Note:

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

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