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Rabbit Anti-Nephrocystin 4 antibody

SL19201R

Product Name:	Nephrocystin 4
Chinese Name:	NPHP4蛋白抗体
Alias:	KIAA0673; Nephrocystin-4; nephronophthisis 4; Nephroretinin; NPHP4; NPHP4_HUMAN; POC10; POC10 centriolar protein homolog; SLSN4.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Sheep,Macaque Monkey, Gorilla, Orangutan
Applications:	ELISA=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:	157kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human Nephrocystin 4:401-500/1426
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:	This gene encodes a protein involved in renal tubular development and function. This protein interacts with nephrocystin, and belongs to a multifunctional complex that is localized to actin- and microtubule-based structures. Mutations in this gene are associated with nephronophthisis type 4, a renal disease, and with Senior-Loken syndrome type 4, a combination of nephronophthisis and retinitis pigmentosa. Alternative splicing results in multiple transcript variants. [provided by RefSeq, Apr

2014]

Function:
Involved in the organization of apical junctions in kidney cells together with NPHP1 and RPGRIP1L/NPHP8. Does not seem to be strictly required for ciliogenesis.

Subcellular Location:
Cytoplasm > cytoskeleton > cilium basal body. Cytoplasm > cytoskeleton > centrosome.

Tissue Specificity:
Expressed in kidney, skeletal muscle, heart and liver, and to a lesser extent in brain and lung.

DISEASE:
Defects in NPHP4 are the cause of nephronophthisis type 4 (NPHP4) [MIM:606966]; also known as familial juvenile nephronophthisis 4. NPHP4 is an autosomal recessive inherited disease resulting in end-stage renal disease at age ranging between 6 and 35 years. It is a progressive tubulo-interstitial kidney disorder characterized by polydipsia, polyuria, anemia and growth retardation. The most prominent histological features are modifications of the tubules with thickening of the basement membrane, interstitial fibrosis and, in the advanced stages, medullary cysts.
Defects in NPHP4 are the cause of Senior-Loken syndrome type 4 (SLSN4) [MIM:606996]. SLSN is a renal-retinal disorder characterized by progressive wasting of the filtering unit of the kidney, with or without medullary cystic renal disease, and progressive eye disease. Typically this disorder becomes apparent during the first year of life.

Similarity:
Belongs to the NPHP4 family.

SWISS:
O75161

Gene ID:
261734

Database links:

Entrez Gene: 261734 Human

Omim: 607215 Human

SwissProt: O75161 Human

Unigene: 462348 Human

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