Product Name: Nephrocystin-4 Rabbit Polyclonal

Antibody

Catalog #: APRab14563



Summary

Production Name Nephrocystin-4 Rabbit Polyclonal Antibody

Description Rabbit Polyclonal Antibody

Host Rabbit

Application IHC-P,IF-P,IF-F,ICC/IF,ELISA

Reactivity Human, Rat, Mouse

Performance

ConjugationUnconjugatedModificationUnmodified

Isotype IgG

Clonality Polyclonal Form Liquid

Storage Store at 4°C short term. Aliquot and store at -20°C long term. Avoid freeze/thaw cycles.

Liquid in PBS containing 50% glycerol, 0.5% protective protein and 0.02% New type

preservative N.

Purification Affinity purification

Immunogen

Buffer

Gene Name NPHP4

Alternative Names NPHP4; KIAA0673; Nephrocystin-4; Nephroretinin

Gene ID 261734.0

O75161. The antiserum was produced against synthesized peptide derived from human **SwissProt ID**

NPHP4. AA range:877-926

Application

Dilution Ratio IHC-P 1:100-1:300, ELISA 1:40000, IF-P/IF-F/ICC/IF 1:50-200

Molecular Weight

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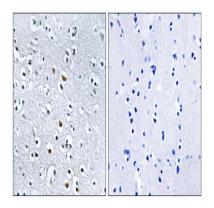


Background

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], 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, disease: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, subunit:Interacts with NPHP1 and RPGRIP1L, tissue specificity:Expressed in kidney, skeletal muscle, heart and liver, and to a lesser extent in brain and lung,

Research Area

Image Data



Immunohistochemistry analysis of paraffin-embedded human brain, using NPHP4 Antibody. The picture on the right is blocked with the synthesized peptide.

Note

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