



Rabbit Anti-Nephrin antibody

SL4866R

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| Product Name: | Nephrin |
| Chinese Name: | 肾小球Cell adhesion molecule受体抗体 |
| Alias: | CNF; Nephrin; Nephrosis 1 congenital Finnish type; Nephrosis 1, congenital, Finnish type (nephrin); NPHN; NPHN_HUMAN; NPHS 1; NPHS1; Renal glomerulus specific cell adhesion receptor; Renal glomerulus-specific cell adhesion receptor. |
| Organism Species: | Rabbit |
| Clonality: | Polyclonal |
| React Species: | Human,Mouse,Rat,Dog,Pig,Cow, |
| 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: | 138kDa |
| Cellular localization: | cytoplasmicThe cell membrane |
| Form: | Lyophilized or Liquid |
| Concentration: | 1mg/ml |
| immunogen: | KLH conjugated synthetic peptide derived from human Nephrin:401-500/1241<Extracellular> |
| 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: | Primary steroid resistant nephrotic syndrome (SRNS) is characterized by childhood onset of proteinuria and progression to end stage renal disease. Kidney podocytes and their slit diaphragms form the final barrier to urinary protein loss. Congenital nephrotic syndrome (CNS) is caused by mutations in NPHS1 (nephrin) or NPHS2. Nephrin, a |

recently identified protein is a member of a group of podocyte proteins that constitute major component of the slit diaphragm especially in the foot process. Nephrin, a cell adhesion molecule, may play a crucial role in maintaining the glomerular filtration barrier. Recent studies have suggested that mutations in the gene for Nephrin reportedly lead to congenital nephrosis. Three novel podocyte proteins, Podocin, Nephrin and alpha Actinin 4 have been identified in congenital and experimental models of proteinuria. The role of Nephrin in anti apoptotic activity in podocyte slit diaphragm is believed to be associated with vascular endothelial derived growth factors VEGF signaling.

Function:

Seems to play a role in the development or function of the kidney glomerular filtration barrier. Regulates glomerular vascular permeability. May anchor the podocyte slit diaphragm to the actin cytoskeleton. Plays a role in skeletal muscle formation through regulation of myoblast fusion.

Subunit:

Interacts with CD2AP (via C-terminal domain). Interacts with MAGI1 (via PDZ 2 and 3 domains) forming a tripartite complex with IGSF5/JAM4. Interacts with DDN; the interaction is direct. Self-associates (via the Ig-like domains). Also interacts (via the Ig-like domains) with KIRREL/NEPH1 and KIRREL2; the interaction with KIRREL is dependent on KIRREL glycosylation. Forms a complex with ACTN4, CASK, IQGAP1, MAGI2, SPTAN1 and SPTBN1. Interacts with NPHS2.

Subcellular Location:

Cell membrane. Predominantly located at podocyte slit diaphragm between podocyte foot processes. Also associated with podocyte apical plasma membrane.

Tissue Specificity:

Specifically expressed in podocytes of kidney glomeruli.

Post-translational modifications:

Phosphorylated at Tyr-1193 by FYN, leading to the recruitment and activation of phospholipase C-gamma-1/PLCG1.

DISEASE:

Defects in NPHS1 are the cause of nephrotic syndrome type 1 (NPHS1) [MIM:256300]; also known as Finnish congenital nephrosis (CNF). A renal disease characterized clinically by proteinuria, hypoalbuminemia, hyperlipidemia, and edema. Kidney biopsies show non-specific histologic changes such as focal segmental glomerulosclerosis and diffuse mesangial proliferation. Some affected individuals have an inherited steroid-resistant form and progress to end-stage renal failure.

Similarity:

Belongs to the immunoglobulin superfamily.
Contains 1 fibronectin type-III domain.

Contains 8 Ig-like C2-type (immunoglobulin-like) domains.

SWISS:

O60500

Gene ID:

4868

Database links:

[Entrez Gene: 4868](#)Human

[Entrez Gene: 54631](#)Mouse

[Entrez Gene: 64563](#)Rat

[Omim: 602716](#)Human

[SwissProt: O60500](#)Human

[SwissProt: Q9QZS7](#)Mouse

[SwissProt: Q9R044](#)Rat

[Unigene: 122186](#)Human

[Unigene: 437830](#)Mouse

[Unigene: 48745](#)Rat

Important Note:

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