



## Rabbit Anti-Nephrin antibody

SL23059R

<b>Product Name:</b>	Nephrin
<b>Chinese Name:</b>	肾小球Cell adhesion molecule受体抗体
<b>Alias:</b>	CNF; NPHN; Nephrosis 1 congenital Finnish type; NPHS 1; NPHS1; Renal glomerulus specific cell adhesion receptor; Renal glomerulus-specific cell adhesion receptor; NPHN_HUMAN
<b>Organism Species:</b>	Rabbit
<b>Clonality:</b>	Polyclonal
<b>React Species:</b>	Human,Mouse,Rat,
<b>Applications:</b>	WB=1:500-2000 not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
<b>Molecular weight:</b>	138kDa
<b>Cellular localization:</b>	cytoplasmicThe cell membrane
<b>Form:</b>	Lyophilized or Liquid
<b>Concentration:</b>	1mg/ml
<b>immunogen:</b>	KLH conjugated synthetic peptide derived from human Nephrin:451-550/1241<Extracellular>
<b>Lsotype:</b>	IgG
<b>Purification:</b>	affinity purified by Protein A
<b>Storage Buffer:</b>	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
<b>Storage:</b>	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.
<b>PubMed:</b>	<a href="#">PubMed</a>
<b>Product Detail:</b>	This gene encodes a member of the immunoglobulin family of cell adhesion molecules that functions in the glomerular filtration barrier in the kidney. The gene is primarily expressed in renal tissues, and the protein is a type-1 transmembrane protein found at the slit diaphragm of glomerular podocytes. The slit diaphragm is thought to function as an ultrafilter to exclude albumin and other plasma macromolecules in the formation of

urine. Mutations in this gene result in Finnish-type congenital nephrosis 1, characterized by severe proteinuria and loss of the slit diaphragm and foot processes.[provided by RefSeq, Oct 2009]

**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 (By similarity). Interacts with NPHS2.

**Subcellular Location:**

Cell membrane; Single-pass type I membrane protein (Potential). Note=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. Expressed in kidney glomeruli. In the embryo, expressed in the mesonephric kidney at E11 with strong expression in cranial tubules with podocyte-like structures. Expression is observed in the podocytes of the developing kidney from E13. High expression is also detected in the developing cerebellum, hindbrain, spinal cord, retina and hypothalamus. Expressed in skeletal muscle during myoblast fusion such as in the adult following acute injury and in the embryo but not detected in uninjured adult skeletal muscle. Isoform 1 and isoform 2 are expressed in the newborn brain and developing cerebellum. Isoform 1 is the predominant isoform in adult kidney.

**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

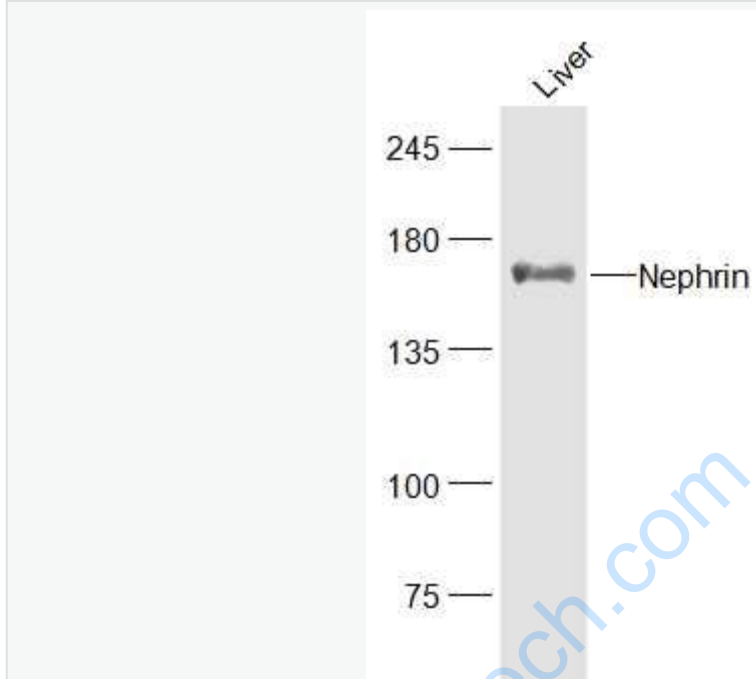
[Unigene: 437830](#)Mouse

[Unigene: 48745](#)Rat

**Important Note:**

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

Picture:



Sample:

Liver (Mouse) Lysate at 40 ug

Primary: Anti-Nephrin (SL23059R) at 1/300 dilution

Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution

Predicted band size: 138 kD

Observed band size: 170 kD