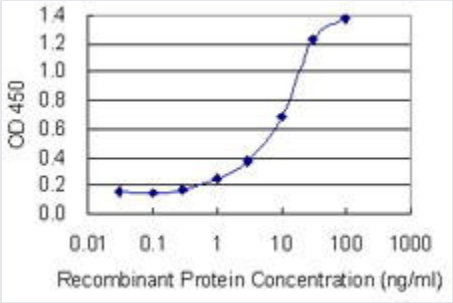
NPHS1 Antibody – Mouse Monoclonal

PRODUCT OVERVIEW

Product Name	NPHS1, Monoclonal Antibody
Full Product Name	NPHS1 (Nephrosis 1, Congenital, Finnish Type (nephrin), CNF, NPHN) (FITC)
Product Gene Name	anti-NPHS1 antibody
Product Synonym Names	Nephrosis 1; Congenital; Finnish Type (nephrin); CNF; NPHN
Clonality	Monoclonal
Isotype	IgG2a,k
Clone Number	3D2
Host	Mouse
Form/Format	Supplied as a liquid in PBS, pH 7.2. No preservative added. Labeled with Fluorescein isothiocyanate (FITC).
Purity/Purification	Purified
Specificity	Recognizes NPHS1.
Preparation and Storage	Store product at 4 degree C if to be used immediately within two weeks. For long-term storage, aliquot to avoid repeated freezing and thawing and store at -20 degree C. Aliquots are stable at -20 degree C for 12 months after receipt. Dilute required amount only prior to immediate use. Further dilutions can be made in assay buffer. FITC conjugates are sensitive to light. For maximum recovery of product, centrifuge the original vial after thawing and prior to removing the cap.
Molecular Weight	130,546 Da
OMIM	256300
3D Structure	ModBase 3D Structure for O60500
Immunogen	NPHS1 (NP_004637.1, 33aa-122aa) partial recombinant protein with GST tag. MW of the GST tag alone is 26kD.
Immunogen Sequence	GFWALPENLTVVEGASVELRCGVSTPGSAVQWAKDGLLLGPDPRIPGFPRYRLEGDPARGEFHLHIEACDLSDDAEYECQVGRSEMGPEL
Conjugate	FITC
Product Categories/Family for anti-NPHS1 antibody	Antibodies; Nephrin
Applications Tested/Suitable for anti-NPHS1 antibody	Western Blot (WB)
Application Notes for anti-NPHS1 antibody	Applications are based on unconjugated antibody.

Related Product Information for anti-NPHS1 antibody	Nephrin is a kidney glomerular filtration barrier protein that is an essential component of the interpodocyte-spanning slit diaphragm. Mutations in the nephrin gene are associated with congenital nephrotic syndrome (NPHS1; MIM 256300). [supplied by OMIM]
Other Notes	Small volumes of anti-NPHS1 antibody vial(s) may occasionally become entrapped in the seal of the product vial during shipment and storage. If necessary, briefly centrifuge the vial on a tabletop centrifuge to dislodge any liquid in the container`s cap. Certain products may require to ship with dry ice and additional dry ice fee may apply.
Searchable Terms for NPHS1 purchase	MBS6178774 is an antibody targeting NPHS1. Also known as NPHS1 (Nephrosis 1, Congenital, Finnish Type (nephrin), CNF, NPHN) (FITC). Host: Mouse.



Testing Data of anti-NPHS1 antibody
Detection limit for recombinant GST tagged NPHS1 is 0.1 ng/ml as a capture antibody.

ADDITIONAL DETAILS

NCBI/Uniprot data below describe general gene information for NPHS1. It may not necessarily be applicable to this product.

NCBI GI #	4758822
NCBI GeneID	4868
NCBI Accession #	NP_004637.1
UniProt Primary Accession #	O60500
UniProt Secondary Accession #	A6NDH2; C3RX61
UniProt Related Accession #	O60500
GenBank Nucleotide #	NM_004646.3
NCBI Official Full Name	nephrin
NCBI Official Synonym Full Names	NPHS1, nephrin
NCBI Official Symbol	NPHS1
NCBI Official Synonym Symbols	CNF; NPHN; nephrin
UniProt Protein Name	Nephrin
UniProt Synonym Protein Names	Renal glomerulus-specific cell adhesion receptor
Protein Family	Nephrin
UniProt Gene Name	NPHS1
UniProt Synonym Gene Names	NPHN
UniProt Entry Name	NPHN_HUMAN
NCBI Protein Information	nephrin

NCBI Summary for NPHS1	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]
UniProt Comments for NPHS1	NPHS1: 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. 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. Specifically expressed in podocytes of kidney glomeruli. Belongs to the immunoglobulin superfamily. 2 isoforms of the human protein are produced by alternative splicing. Protein type: Immunoglobulin superfamily; Cell adhesion; Membrane protein, integral Chromosomal Location of Human Ortholog: 19q13.1 Cellular Component: cell projection; protein complex; integral to plasma membrane; plasma membrane; lipid raft Molecular Function: protein domain specific binding; protein binding; myosin binding; spectrin binding; alpha-actinin binding Biological Process: skeletal muscle development; positive regulation of actin filament polymerization; glomerular basement membrane development; JNK cascade; excretion; cell adhesion; myoblast fusion Disease: Nephrotic Syndrome, Type 1
Research Articles on NPHS1	**1.** The classical form is CNF, which is caused by mutations in the nephrin gene (NPHS1), leading to massive proteinuria, hypoproteinemia and edema in the newborn period
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