IBL Data Sheet

Code No. 29050

Anti-Human
Nephrin (N) Rabbit IgG Affinity Purify

Volume : 100 µg
Lot No. :

Introduction : The kidney is an important organ that maintains a homeostasis of body-fluid and nutrition. The capillary walls of renal glomeruli allow the efficient removal of the metabolic waste (filter) and conservation of essential circulating proteins, such as albumin (barrier). The filtration barriers are composed of three layers: a fenestrated endothelium, the glomerular basement membrane, and the highly specialized epithelial cells, podocytes. The podocytes elaborate numerous cellular processes and the terminal portions, called foot process, cover the outmost surface of the glomerular basement membrane. The neighboring foot processes are aligned in an interdigitating fashion while leaving a filtration slit around 20-50nm in wide that is bridged by an electron dense membrane-like structure, the slit diaphragm. Recent studies with familial nephrotic syndrome and genetically manipulated mice models demonstrated the genetic defects of the slit diaphragm proteins (nephrin, podocin, etc) cause massive proteinuria (nephrotic syndrome) and eventually progress the end stage renal diseases. The observations indicate that regulation of the slit diaphragm integrity is crucial for maintenance of the filtration barrier function and podocyte viability.

Nephrin has been identified as a disease causing molecule responsible for congenital nephrotic syndrome of Finnish type, in which the proteinuria starts *in utero* or immediately after birth (ref. 1). It is an immunoglobulin-like transmembrane adhesion protein having eight Ig motifs. Nephrin forms a structural backbone of the slit diaphragm through a head-to-head, trans-homo-interaction at the N-terminal ectodomains. In addition to the structural function, nephrin serves as a signaling molecule that mediate an adaptive morphological regulation through the remodeling of actin cytoskeletons; the C-terminal cytoplasmic domain is tyrosine-phosphorylated by Src family kinase (Fyn) and the phosphorylated nephrin associates with an SH2 adaptor protein (Nck), thereby allowing the sable anchorage of the slit membrane complex to the actin cytoskeleton in the foot processes. Under proteinuric conditions, nephrin is down-regulated and is apically dislocated (ref. 2).

Antigen : Recombinant protein of Human Nephrin

Purification : Purified with antigen

Form : Lyophilized product from 1% BSA in PBS containing 0.05% NaN_3

How to use : Add 1.0 mL deionized water into the product (the conc. comes up 100 µg /mL).

Stability :

Lyophilized product	5 years at 2 - 8 °C
Solution	2 years at –20 °C

Application : The recommended dilutions is as follow but the optimal concentration should be determined by researchers.
: For immunohistochemistry in a frozen section, the starting concentration is 5 µg/mL. (mainly can be used for study of cell surface)
: Western blotting (W.B.) in concentration of 1 µg /mL

Reference :

1. Kestilä M, Lenkkeri U, Männikkö M, Lamerdin J, McCready P, Putaala H, Ruotsalainen V, Morita T, Nissinen M, Herva R, Kashtan CE, Peltonen L, Holmberg C, Olsen A, Tryggvason K. Positionally cloned gene for a novel glomerular protein--nephrin--is mutated in congenital nephrotic syndrome.Mol Cell. 1998 Mar;1(4):575-82.
2. Kitamura A, Tsukaguchi H, Hiramoto R, Shono A, Doi T, Kagami S, Iijima K. A familial childhood-onset relapsing nephrotic syndrome.Kidney Int. 2007 May;71(9):946-51.

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