

DESCRIPTION

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| Species Reactivity | Human |
| Specificity | Detects human Norrin in Western blots. In Western blots, approximately 50% cross-reactivity with recombinant mouse Norrin is observed. |
| Source | Polyclonal Goat IgG |
| Purification | Antigen Affinity-purified |
| Immunogen | <i>E. coli</i> -derived recombinant human Norrin Lys25-Ser133 Accession # Q00604 |
| Formulation | Lyophilized from a 0.2 µm filtered solution in PBS with BSA as a carrier protein. |

APPLICATIONS

Please Note: Optimal dilutions should be determined by each laboratory for each application. [General Protocols](#) are available in the [Technical Information](#) section on our website.

| | Recommended Concentration | Sample |
|---------------------|---------------------------|--|
| Western Blot | 0.1 µg/mL | Recombinant Human Norrin (Catalog # 3014-NR) |

PREPARATION AND STORAGE

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|--------------------------------|---|
| Reconstitution | Reconstitute at 0.2 mg/mL in sterile PBS. |
| Shipping | The product is shipped at ambient temperature. Upon receipt, store it immediately at the temperature recommended below. |
| Stability & Storage | <p>Use a manual defrost freezer and avoid repeated freeze-thaw cycles.</p> <ul style="list-style-type: none"> • 12 months from date of receipt, -20 to -70 °C as supplied. • 1 month, 2 to 8 °C under sterile conditions after reconstitution. • 6 months, -20 to -70 °C under sterile conditions after reconstitution. |

BACKGROUND

Norrin (also called Norrie Disease Protein or NDP) is a secreted regulatory protein that remains tightly associated with the extracellular matrix (1). At least 70 mutations of Norrin have been identified in Norrie disease (X-linked retinal dysplasia) or in a minority of X-linked familial exudative vitreoretinopathy (FEVR). Both are disorders of retinal vascularization that cause blindness (2, 3). Norrin consists of disulfide-linked homodimers that oligomerize further via disulfide bridges to form higher order oligomers containing up to ten units of 12 kDa each. The cysteine-rich C-terminal domain of Norrin is homologous to von Willebrand factor, several extracellular mucin proteins, and members of the TGF-β family. Molecular modeling studies predict that Norrin assumes a cysteine-knot structure typical for the TGF-β family (4). Although Norrin is not related to Wnt family proteins, it functions like a Wnt protein in that it binds with high affinity to the receptor Frizzled-4, requires LDL receptor-related protein (LRP) as a co-receptor, and induces activation of the canonical Wnt signaling pathway (5). Norrin and Frizzled-4 are expressed at relatively low levels in tissues displaying vascular phenotypes and genetic disruption of either gene in mice gives phenotypes with marked similarities (5, 6, 7). Human Norrin shares 100%, 96%, 95%, 94% and 90% amino acid identity with rhesus macaque, canine, bovine, mouse, and chick Norrin, respectively.

References:

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3. Berger, W. and H.H. Ropers (2001) "*The Metabolic and Molecular Bases of Inherited Diseases*", C.R. Scriver, *et al.* eds. p. 5977.
4. Meitinger, T *et al.* (1993) *Nat Genet* **5**:376.
5. Xu, Q. *et al.* (2004) *Cell* **116**:883.
6. Hartzler, M.K. *et al.* (1999) *Brain Res. Bull.* **49**:355.
7. Wang, Y. *et al.* (2001) *J. Neurosci.* **21**:4761.