Product Informatiion Sheet

Monoclonal Anti-Nebulin (Sepharose Bead Conjugate)

Catalogue No. MA1069-S

Lot No. 08A12

Clone: Neb-20

Ig type: mouse IgG1

Size: 200µl

Specificity

Human, mouse, rat, chicken, frog.
No cross reactivity with other proteins.

Recommended application

Immunoprecipitation(IP)

Immunogen

Chicken breast nebulin.

Purification

Purified by the goat anti-mouse IgG affinity chromatography.

Formulation

50% slurry in PBS pH 7.2 with 0.01mg NaN3a3 preservative.

Storage

Store at 4˚C for frequent use.

Description:

This Antagene antibody is immobilized via covalent binding of primary amino groups to N-hydroxysuccinimide (NHS)-activated sepharose beads. It is useful for immunoprecipitation assays

BACKGROUND

Nebulin is a giant protein component of the cytoskeletal matrix that coexists with the thick and thin filaments within the sarcomeres of skeletal muscle. In most vertebrates, nebulin accounts for 3 to 4% of the total myofibrillar protein and its size varies from 600 to 800 kD in a manner that is tissue-, species-, and developmental stage-specific. The nebulin gene contains 183 exons in a 249-kb genomic region. Nebulin is mapped to chromosome 2. Nebulin is a giant filamentous protein specific for vertebrate skeletal muscles. Mutations in the nebulin gene associated with autosomal recessive nemaline myopathy.

REFERENCE

1. Limongi, M. Z.; Pelliccia, F.; Rocchi, A. : Assignment of the human nebulin gene (NEB) to chromosome band 2q24.2 and the alpha-1 (III) collagen gene (COL3A1) to chromosome band 2q32.2 by in situ hybridization: the FRA2G common fragile site lies between the two genes in the 2q31 band. *Cytogenet. Cell Genet.* 77: 259-260, 1997.

2. Labeit, S.; Kolmerer, B. : The complete primary structure of human nebulin and its correlation to muscle structure. *J. Molec. Biol.* 248: 308-315, 1995. 3. Pelin, K.; Hilpela, P.; Donner, K.; Sewry, C.; Akkari, P. A.; Wilton, S. D.; Wattanasirichaigoon, D.; Bang, M.-L.; Centner, T.; Hanefeld, F.; Odent, S.; Fardeau, M.; Urtizberea, J. A.; Muntoni, F.; Dubowitz, V.; Beggs, A. H.; Laing, N. G.; Labeit, S.; de la Chapelle, A.; Wallgren-Pettersson, C. : Mutations in the nebulin gene associated with autosomal recessive nemaline myopathy. *Proc. Nat. Acad. Sci.* 96: 2305-2310, 1999.