

# Monoclonal Anti- Nebulin Antibody

Catalog Number: MA1069

## Description

Lot No.	08A12
Clone	Neb-20
Size	100µg/vial
Form	lyophilized
Ig type	mouse IgG1
Specificity	No cross reactivity with other proteins.
Species	Chicken
Immunogen	Chicken breast nebulin.
Contents	Mouse ascites fluid, 1.2% sodium acetate, 2mg BSA, with 0.01mg NaN <sub>3</sub> as preservative.

## Application

Western blot At 1-2 µg/ml with the appropriate system to detect nebulin in cells and tissues.

*Other applications have not been tested.*

*Optimal dilutions should be determined by end users.*

## Preparation and storage

**Reconstitution:** 1.2% sodium acetate or neutral PBS. If 1ml of PBS is used, the antibody concentration will be 100µg/ml.

**Storage:** At -20°C for one year. After reconstitution, at 4°C for one month. It can also be aliquotted and stored frozen at -20°C for a longer time.

Avoid repeated freezing and thawing.

## Relevant detection systems

We provide a series of assays reacted with primary antibodies. Antibody can be supported by chemiluminescence kit EK1001 in WB.

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## 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.