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ProductInformation

Monoclonal Anti-MBNL1
Clone HL 1822 (3A4-1E9)
produced in mouse, purified immunoglobulin

Catalog Number **M3320**

Product Description

Monoclonal Anti-MBNL1 (mouse IgG1 isotype) is derived from the hybridoma HL1822 (3A4-1E9) produced by the fusion of mouse myeloma cells (SP2/0 cells) and splenocytes from BALB/c mice immunized with recombinant Muscle Blind-Like1 (MBNL1) fusion protein. The isotype is determined using a double diffusion immunoassay using Mouse Monoclonal Antibody Isotyping Reagents, Catalog Number ISO2.

Monoclonal Anti-MBNL1 recognizes human,[1, 2] monkey,[3] and mouse[4] MBNL1. The antibody may used in immunoblotting (~42 kDa),[1, 4] immunohistochemistry,[2] and immunocytochemistry.[1, 3, 4]

Myotonic dystrophy (DM-dystrophia myotonica), a neuromuscular disease, is caused by microsatellite repeat expansions at two different genomic loci. This disease is characterized by myotonia or delayed muscle relaxation due to repetitive action potentials in myofibers and muscle degeneration. Expansion of cytosine-thymine-guanine (CTG)n repeats in the 3'-untranslated region of DM protein kinase DMPK gene on chromosome 19, is responsible for DM type 1, while expansions of cytosine-cytosine-thymine-guanine (CCTG)n in the first intron of the zinc finger protein 9 (ZNF9) gene is responsible for DM type 2. Transcripts of these genes accumulate in foci within muscle nuclei. Proteins of the MBNL (muscleblind-like) family are colocalized with mutant DM transcripts and bind to CUG repeats *in vitro*.[1-4]

Human muscle blind homologs MBNL1, MBNL2 and MBNL3 promote inclusion or exclusion of specific exons on different pre-mRNAs by antagonizing the activity of CUG-BP and ETR-3-like factors (CELF proteins) bound to distinct intronic sites. MBNL1 seems to promote muscle differentiation while MBNL3 seems to inhibit the expression of muscle differentiation markers. MBNL2 participates in a new RNA-dependent protein localization mechanism involving recruitment of integrin α3 protein to focal adhesions.[5] The muscle blind family of proteins is expressed in skeletal muscle, and has multiple protein isoforms, including some that bind to expanded CUG repeats (41-42 kDa) while others that fail to bind (31 kDa). Mice knockouts for the *MBNL1* gene develop muscle, eye, and RNA splicing abnormalities that are characteristic of the DM disease.[4]

Reagent

Supplied as a solution in 0.01 M phosphate buffered saline, pH 7.4, containing 15 mM sodium azide.

Antibody concentration: ~1 mg/mL

Precautions and Disclaimer

This product is for R&D use only, not for drug, household, or other uses. Please consult the Material Safety Data Sheet for information regarding hazards and safe handling practices.

Storage/Stability

For continuous use, store at 2-8 °C for up to one month. For extended storage, freeze in working aliquots. Repeated freezing and thawing, or storage in "frost-free" freezers, is not recommended. If slight turbidity occurs upon prolonged storage, clarify the solution by centrifugation before use. Working dilution samples should be discarded if not used within 12 hours.

Product Profile

Immunoblotting: a working concentration of 0.2-0.5 µg/mL is recommended using HeLa nuclear cell extract.

Note: In order to obtain the best results using various techniques and preparations, we recommend determining optimal working dilutions by titration.

References

1. Ho, T.H., et al., *EMBO J.*, **23**, 3103-3112 (2004).
2. Jiang, H., et al., *Hum. Mol. Gen.*, **13**, 3079-3088 (2004).
3. Ho, T.H., et al., *J. Cell Sci.*, **118**, 2923-2933 (2005).

4. Kanadia, R.N., et al., *Science*, **302**, 1978-1980 (2003)
5. Pascual, M., *Differentiation*, **74**, 65-80 (2006).