

MBNL1/2/3 (H-90): sc-98455

BACKGROUND

Pre-mRNA splicing is a critical step in the posttranscriptional regulation of gene expression. Several protein complexes are involved in proper mRNA splicing and transport. The muscleblind proteins, 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 bound to distinct intronic sites. MBNL1 is a deduced 370 amino acid protein which is predominantly expressed in skeletal muscle, prostate, lung, heart, small intestine, ovary and placenta tissues. MBNL1 and MBNL2, which associate with expanded CUG repeats *in vitro*, both localize to the nuclear foci in both DM1 and DM2 (myotonic dystrophy types 1 and 2), suggesting that the nuclear accumulation of mutant RNA is pathogenic in DM1, therefore implicating MBNL1 and 2 in the pathogenesis of both disorders.

REFERENCES

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3. Mankodi, A., et al. 2001. Muscleblind localizes to nuclear foci of aberrant RNA in myotonic dystrophy types 1 and 2. Hum. Mol. Genet. 10: 2165-2170.
4. Fardaei, M., et al. 2002. Three proteins, MBNL, MBLL and MBXL, co-localize *in vivo* with nuclear foci of expanded-repeat transcripts in DM1 and DM2 cells. Hum. Mol. Genet. 11: 805-814.
5. Ho, T.H., et al. 2005. Co-localization of muscleblind with RNA foci is separable from mis-regulation of alternative splicing in myotonic dystrophy. J. Cell Sci. 118: 2923-2933.
6. Ladd, A.N., et al. 2005. Dynamic balance between activation and repression regulates pre-mRNA alternative splicing during heart development. Dev. Dyn. 233: 783-793.
7. Ishiura, S., et al. 2005. Molecular pathways to myotonic dystrophy. Nippon Rinsho 63: 515-521.
8. Dansithong, W., et al. 2005. MBNL1 is the primary determinant of focus formation and aberrant Insulin receptor splicing in DM1. J. Biol. Chem. 280: 5773-5780.

SOURCE

MBNL1/2/3 (H-90) is a rabbit polyclonal antibody raised against amino acids 11-100 mapping near the N-terminus of MBNL1 of human origin.

PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

STORAGE

Store at 4° C, ****DO NOT FREEZE****. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

APPLICATIONS

MBNL1/2/3 (H-90) is recommended for detection of MBNL1/2/3 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

MBNL1/2/3 (H-90) is also recommended for detection of MBNL1/2/3 in additional species, including equine, canine, bovine, porcine and avian.

Molecular Weight of MBNL1: 42 kDa.

Molecular Weight of MBNL2: 41 kDa.

Molecular Weight of MBNL3: 39 kDa.

Positive Controls: HeLa whole cell lysate: sc-2200.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

RESEARCH USE

For research use only, not for use in diagnostic procedures.

PROTOCOLS

See our web site at www.scbt.com or our catalog for detailed protocols and support products.



Try **MBNL1 (D-4): sc-515374** or **MBNL1/2/3 (MBNL): sc-58790**, our highly recommended monoclonal alternatives to MBNL1/2/3 (H-90).