SANTA CRUZ BIOTECHNOLOGY, INC.

MBNL1/2/3 (MBNL): sc-58790

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The Power to Question

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

1. Ishikawa, K., et al. 1998. Prediction of the coding sequences of unidentified human genes. VIII. 78 new cDNA clones from brain which code for large proteins *in vitro.* DNA Res. 4: 307-313.
2. Miller, J.W., et al. 2000. Recruitment of human muscleblind proteins to (CUG)(n) expansions associated with myotonic dystrophy. EMBO J. 19: 4439-4448.
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. Colocalization 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 (MBNL) is a mouse monoclonal antibody raised against a 371 amino acid fragment of MBNL1 of human origin.

PRODUCT

Each vial contains 250 µl culture supernatant containing IgG_1 with < 0.1% sodium azide.

RESEARCH USE

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

APPLICATIONS

MBNL1/2/3 (MBNL) is recommended for detection of MBNL1/2/3 of mouse, rat and human origin by Western Blotting (starting dilution to be determined by researcher, dilution range 1:500-1:2500), immunoprecipitation [1-2 µl per 100-500 µg of total protein (1 ml of cell lysate)] and immunofluorescence (starting dilution to be determined by researcher, dilution range 1:10-1:200).

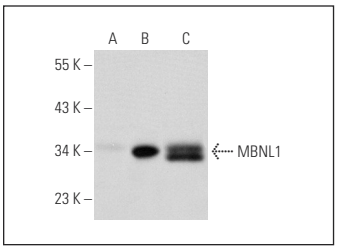
Molecular Weight of MBNL1 isoforms: 42 kDa.

Molecular Weight of MBNL2: 40.5 kDa.

Molecular Weight of MBNL3: 39 kDa.

Positive Controls: MBNL1 (h): 293T Lysate: sc-115973 or HeLa whole cell lysate: sc-2200.

DATA



MBNL1/2/3 (MBNL): sc-58790. Western blot analysis of MBNL1 expression in non-transfected 293T: sc-117752 (**A**), human MBNL1 transfected 293T: sc-115973 (**B**) and HeLa (**C**) whole cell lysates.

STORAGE

For immediate and continuous use, store at 4° C for up to one month. For sporadic use, freeze in working aliquots in order to avoid repeated freeze/thaw cycles. If turbidity is evident upon prolonged storage, clarify solution by centrifugation.

CONJUGATES

See **MBNL2 (3B4): sc-136167** for MBNL2 antibody conjugates, including AC, HRP, FITC, PE, Alexa Fluor® 488 and Alexa Fluor® 647.