SANTA CRUZ BIOTECHNOLOGY, INC.

MBNL3 (5A11): sc-136168

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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 and 2, which associate with expanded CUG repeats *in vitro,* 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. MBNL3, a 354 amino acid protein, inhibits expression of muscle differentiation, opposite to the function of MBNL1, which functions as a promoter of muscle differentiation. MBNL3 shows strong expression in placenta.

REFERENCES

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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. 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.
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CHROMOSOMAL LOCATION

Genetic locus: MBNL3 (human) mapping to Xq26.2; Mbnl3 (mouse) mapping to X A5.

SOURCE

MBNL3 (5A11) is a mouse monoclonal antibody raised against recombinant MBNL3 of human origin.

PRODUCT

Each vial contains 200 µg IgG_1 kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

MBNL3 (5A11) is recommended for detection of MBNL3 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and immunohistochemistry (including paraffin-embedded sections) (starting dilution 1:50, dilution range 1:50-1:500).

Suitable for use as control antibody for MBNL3 siRNA (h): sc-60992, MBNL3 siRNA (m): sc-60993, MBNL3 shRNA Plasmid (h): sc-60992-SH, MBNL3 shRNA Plasmid (m): sc-60993-SH, MBNL3 shRNA (h) Lentiviral Particles: sc-60992-V and MBNL3 shRNA (m) Lentiviral Particles: sc-60993-V.

Molecular Weight of MBNL3: 39 kDa.

RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunofluorescence: use m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850. 3) Immunohistochemistry: use m-IgGκ BP-HRP: sc-516102 with DAB, 50X: sc-24982 and Immunohistomount: sc-45086, or Organo/Limonene Mount: sc-45087.

SELECT PRODUCT CITATIONS

1. Mérien, A., et al. 2021. CRISPR gene editing in pluripotent stem cells reveals the function of MBNL proteins during human *in vitro* myogenesis. Hum. Mol. Genet. E-published.

STORAGE

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

RESEARCH USE

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

PROTOCOLS

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