

## MBNL2 (D-1): sc-365104



The Power to Question

## BACKGROUND

Pre-mRNA splicing is a critical step in the post-transcriptional 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 MBNL2, associate with expanded CUG repeats *in vitro* and 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 muscleblind proteins MBNL1 and MBNL2 in the pathogenesis of both disorders. MBNL2, a 367 amino acid protein, participates in recruitment of Intergrin  $\alpha 3$  to focal adhesions in a RNA-dependent protein localization mechanism.

## REFERENCES

1. Ishikawa, K., et al. 1997. 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)<sub>n</sub> 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 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.

## CHROMOSOMAL LOCATION

Genetic locus: MBNL2 (human) mapping to 13q32.1; Mbnl2 (mouse) mapping to 14 E4.

## SOURCE

MBNL2 (D-1) is a mouse monoclonal antibody raised against amino acids 101-181 mapping within an internal region of MBNL2 of human origin.

## PRODUCT

Each vial contains 200  $\mu$ g IgG<sub>1</sub> kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

## APPLICATIONS

MBNL2 (D-1) is recommended for detection of MBNL2 of mouse, rat and human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2  $\mu$ g per 100-500  $\mu$ g 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).

Suitable for use as control antibody for MBNL2 siRNA (h): sc-60990, MBNL2 siRNA (m): sc-60991, MBNL2 shRNA Plasmid (h): sc-60990-SH, MBNL2 shRNA Plasmid (m): sc-60991-SH, MBNL2 shRNA (h) Lentiviral Particles: sc-60990-V and MBNL2 shRNA (m) Lentiviral Particles: sc-60991-V.

Molecular Weight of MBNL2: 41 kDa.

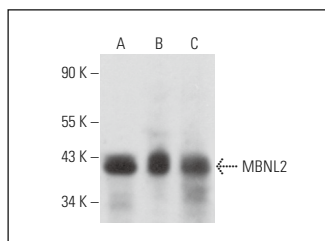
Positive Controls: HeLa whole cell lysate: sc-2200, rat brain extract: sc-2392 or mouse brain extract: sc-2253.

## RECOMMENDED SUPPORT REAGENTS

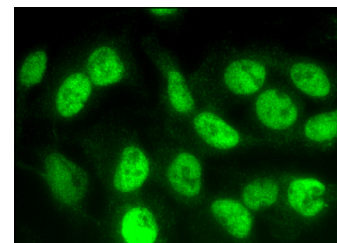
To ensure optimal results, the following support reagents are recommended:

- 1) Western Blotting: use m-IgG $\kappa$  BP-HRP: sc-516102 or m-IgG $\kappa$  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) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml).
- 3) Immunofluorescence: use m-IgG $\kappa$  BP-FITC: sc-516140 or m-IgG $\kappa$  BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

## DATA



MBNL2 (D-1): sc-365104. Western blot analysis of MBNL2 expression in HeLa whole cell lysate (A) and rat brain (B) and mouse brain (C) tissue extracts.



MBNL2 (D-1): sc-365104. Immunofluorescence staining of formalin-fixed Hep G2 cells showing nuclear localization.

## 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.



See **MBNL2 (3B4): sc-136167** for MBNL2 antibody conjugates, including AC, HRP, FITC, PE, and Alexa Fluor® 488, 546, 594, 647, 680 and 790.