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- Mindermengenzuschlag
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SANTA CRUZ BIOTECHNOLOGY, INC.

MBNL1 (h): 293T Lysate: sc-115973

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 distince 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 muscleblind proteins 1 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* wit 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.

CHROMOSOMAL LOCATION

Genetic locus: MBNL1 (human) mapping to 3q25.1.

PRODUCT

MBNL1 (h): 293T Lysate represents a lysate of human MBNL1 transfected 293T cells and is provided as 100 µg protein in 200 µl SDS-PAGE buffer.

STORAGE

Store at -20° C. Repeated freezing and thawing should be minimized. Sample vial should be boiled once prior to use. Non-hazardous. No MSDS required.

APPLICATIONS

MBNL1 (h): 293T Lysate is suitable as a Western Blotting positive control for human reactive MBNL1 antibodies. Recommended use: 10-20 µl per lane.

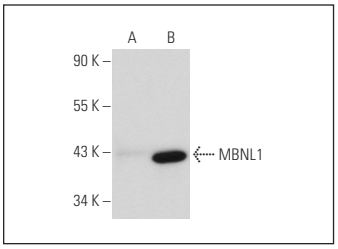
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

MBNL1 (3A4): sc-47740 is recommended as a positive control antibody for Western Blot analysis of enhanced human MBNL1 expression in MBNL1 transfected 293T cells (starting dilution 1:100, dilution range 1:100-1:1,000).

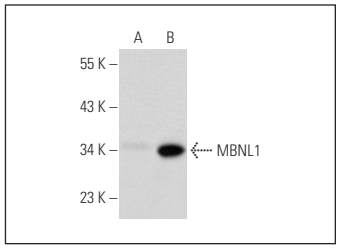
RECOMMENDED SUPPORT REAGENTS

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.

DATA



MBNL1 (3A4): sc-47740. Western blot analysis of MBNL1 expression in non-transfected: sc-117752 (**A**) and human MBNL1 transfected: sc-115973 (**B**) 293T whole cell lysates.



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

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.