SANTA CRUZ BIOTECHNOLOGY, INC.

MBNL1 (3A4): sc-47740

SANTA CRUZ BIOTECHNOLOGY

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 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.

CHROMOSOMAL LOCATION

Genetic locus: MBNL1 (human) mapping to 3q25.1; Mbnl1 (mouse) mapping to 3 D.

SOURCE

MBNL1 (3A4) is a mouse monoclonal antibody raised against recombinant MBNL1 fusion protein 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.

MBNL1 (3A4) is available conjugated to agarose (sc-47740 AC), 500 µg/ 0.25 ml agarose in 1 ml, for IP; to HRP (sc-47740 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-47740 PE), fluorescein (sc-47740 FITC), Alexa Fluor® 488 (sc-47740 AF488), Alexa Fluor® 546 (sc-47740 AF546), Alexa Fluor® 594 (sc-47740 AF594) or Alexa Fluor® 647 (sc-47740 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-47740 AF680) or Alexa Fluor® 790 (sc-47740 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

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APPLICATIONS

MBNL1 (3A4) is recommended for detection of MBNL1 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 immunohistochemistry (including paraffin-embedded sections) (starting dilution 1:50, dilution range 1:50-1:500).

Suitable for use as control antibody for MBNL1 siRNA (h): sc-60988, MBNL1 siRNA (m): sc-60989, MBNL1 shRNA Plasmid (h): sc-60988-SH, MBNL1 shRNA Plasmid (m): sc-60989-SH, MBNL1 shRNA (h) Lentiviral Particles: sc-60988-V and MBNL1 shRNA (m) Lentiviral Particles: sc-60989-V.

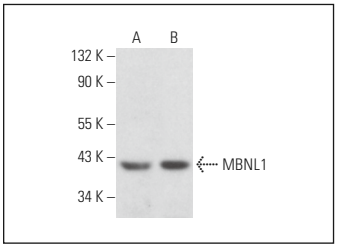
Molecular Weight of MBNL1: 42 kDa.

Positive Controls: MBNL1 (h): 293T Lysate: sc-115973, THP-1 cell lysate: sc-2238 or RAW 264.7 whole cell lysate: sc-2211.

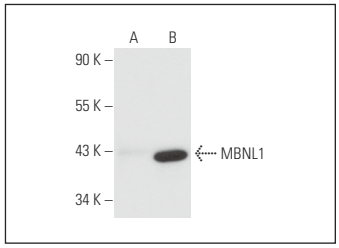
STORAGE

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

DATA



MBNL1 (3A4): sc-47740. Western blot analysis of MBNL1 expression in THP-1 (**A**) and RAW 264.7 (**B**) whole cell lysates.



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.

SELECT PRODUCT CITATIONS

1. Sen, S., et al. 2010. Muscleblind-like 1 (MBNL1) promotes Insulin receptor exon 11 inclusion via binding to a downstream evolutionarily conserved intronic enhancer. J. Biol. Chem. 285: 25426-25437.
2. Hsu R.J., et al. 2011. Long tract of untranslated CAG repeats is deleterious in transgenic mice. PLoS ONE 6: e16417.
3. Laurent, F.X., et al. 2012. New function for the RNA helicase p68/DDX5 as a modifier of MBNL1 activity on expanded CUG repeats. Nucleic Acids Res. 40: 3159-3171.
4. Lee, Y.B., et al. 2013. Hexanucleotide repeats in ALS/FTD form length-dependent RNA foci, sequester RNA binding proteins, and are neurotoxic. Cell Rep. 5: 1178-1186.
5. Cheng, A.W., et al. 2014. Muscleblind-like 1 (MBNL1) regulates pre-mRNA alternative splicing during terminal erythropoiesis. Blood 124: 598-610.
6. Du, J., et al. 2015. RNA toxicity and missplicing in the common eye disease fuchs endothelial corneal dystrophy. J. Biol. Chem. 290: 5979-5990.
7. Alves, S., et al. 2016. Lentiviral vector-mediated overexpression of mutant ataxin-7 recapitulates SCA7 pathology and promotes accumulation of the FUS/TLS and MBNL1 RNA-binding proteins. Mol. Neurodegener. 11: 58.
8. Provenzano, C., et al. 2017. CRISPR/Cas9-mediated deletion of CTG expansions recovers normal phenotype in myogenic cells derived from myotonic dystrophy 1 patients. Mol. Ther. Nucleic Acids 9: 337-348.
9. Morriss, G.R., et al. 2018. Mechanisms of skeletal muscle wasting in a mouse model for myotonic dystrophy type 1. Hum. Mol. Genet. 27: 2789-2804.

RESEARCH USE

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