

CUG-BP1 (m2): 293T Lysate: sc-126681

BACKGROUND

Myotonic dystrophy (DM) is an autosomal dominant neuromuscular disease that is associated with a (CTG)_n repeat expansion in the 3'-untranslated region of the myotonin protein kinase gene (DMPK). CUG-BP1 and CUG-BP2 are proteins that bind specifically to (CUG)₈ oligonucleotides *in vitro*. While CUG-BP1 has the major binding activity in normal cells, nuclear CUG-BP2 binding activity increases in DM cells. Both CUG-BP1 and CUG-BP2 are isoforms of a novel heterogeneous nuclear ribonucleoprotein (hnRNP), hnAB50. CUG-BP1, an RNA CUG triplet repeat binding protein, regulates splicing and translation of various RNAs. Expansion of RNA CUG repeats in the DMPK in DM is associated with alterations in binding activity of CUG-BP1 as well as alterations in the translation of the C/EBP β transcription factor. CUG-BP1 is an important regulator of initiation from different AUG codons of C/EBP β mRNA. In normal cells, CUG-BP1 up-regulates the p21 protein during differentiation by inducing the translation of p21 via binding to a GC-rich sequence located within the 5' region of p21 mRNA. In DM cells, failure to accumulate CUG-BP1 leads to a reduction of p21 and alterations in other proteins responsible for cell cycle withdrawal.

REFERENCES

1. Timchenko, L.T., Miller, J.W., Timchenko, N.A., DeVore, D.R., Datar, K.V., Liu, L., Roberts, R., Caskey, C.T. and Swanson, M.S. 1996. Identification of a (CUG)_n triplet repeat RNA-binding protein and its expression in myotonic dystrophy. *Nucleic Acids Res.* 24: 4407-4414.
2. Timchenko, N.A., Welm, A.L., Lu, X. and Timchenko, L.T. 1999. CUG repeat binding protein (CUGBP1) interacts with the 5'-region of C/EBP β mRNA and regulates translation of C/EBP β isoforms. *Nucleic Acids Res.* 27: 4517-4525.
3. Takahashi, N., Sasagawa, N., Suzuki, K. and Ishiura, S. 2000. The CUG-binding protein binds specifically to UG dinucleotide repeats in yeast three-hybrid system. *Biochem. Biophys. Res. Commun.* 277: 518-523.
4. Timchenko, N.A., Cai, Z.J., Welm, A.L., Reddy, S., Ashizawa, T. and Timchenko, L.T. 2001. RNA CUG repeats sequester CUG-BP1 and alter protein levels and activity of CUG-BP1. *J. Biol. Chem.* 276: 7820-7826.
5. Timchenko, N.A., Iakova, P., Cai, Z.J., Smith, J.R. and Timchenko, L.T. 2001. Molecular basis for impaired muscle differentiation in myotonic dystrophy. *Mol. Cell. Biol.* 21: 6927-6938.

CHROMOSOMAL LOCATION

Genetic locus: Cugbp1 (mouse) mapping to 2 E1.

PRODUCT

CUG-BP1 (m2): 293T Lysate represents a lysate of mouse CUG-BP1 transfected 293T cells and is provided as 100 μ g protein in 200 μ l 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.

RESEARCH USE

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

APPLICATIONS

CUG-BP1 (m2): 293T Lysate is suitable as a Western Blotting positive control for mouse reactive CUG-BP1 antibodies. Recommended use: 10-20 μ l per lane.

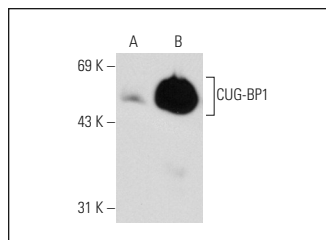
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

CUG-BP1/2 (B-1): sc-166095 is recommended as a positive control antibody for Western Blot analysis of enhanced mouse CUG-BP1 expression in CUG-BP1 transfected 293T cells (starting dilution 1:100, dilution range 1:100-1:1,000).

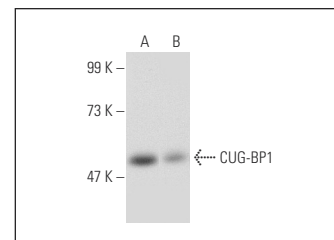
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended:
1) Western Blotting: use m-IgG κ BP-HRP: sc-516102 or m-IgG κ 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



CUG-BP1/2 (B-1): sc-166095. Western blot analysis of CUG-BP1 expression in non-transfected: sc-117752 (A) and mouse CUG-BP1 transfected: sc-126681 (B) 293T whole cell lysates.



CUG-BP1 (3B1): sc-20003. Western blot analysis of CUG-BP1 expression in non-transfected: sc-117752 (A) and mouse CUG-BP1 transfected: sc-126680 (B) 293T whole cell lysates.

PROTOCOLS

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