

CUG-BP1/2 (H-186): sc-48822

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., et al. 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., et al. 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., et al. 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., et al. 2001. RNA CUG repeats sequester CUGBP1 and alter protein levels and activity of CUGBP1. *J. Biol. Chem.* 276: 7820-7826.
5. Timchenko, N.A., et al. 2001. Molecular basis for impaired muscle differentiation in myotonic dystrophy. *Mol. Cell. Biol.* 21: 6927-6938.

CHROMOSOMAL LOCATION

Genetic locus: CELF1 (human) mapping to 11p11.2, CELF2 (human) mapping to 10p14; Celf1 (mouse) mapping to 2 E1, Celf2 (mouse) mapping to 2 A1.

SOURCE

CUG-BP1/2 (H-186) is a rabbit polyclonal antibody raised against amino acids 9-194 mapping near the N-terminus of CUG-BP1 of human origin.

PRODUCT

Each vial contains 200 μ g IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

STORAGE

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

APPLICATIONS

CUG-BP1/2 (H-186) is recommended for detection of CUG-BP1 and CUG-BP2 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 μ g per 100-500 μ 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).

CUG-BP1/2 (H-186) is also recommended for detection of CUG-BP1 and CUG-BP2 in additional species, including equine, canine, bovine and avian.

Molecular Weight of CUG-BP1: 56 kDa.

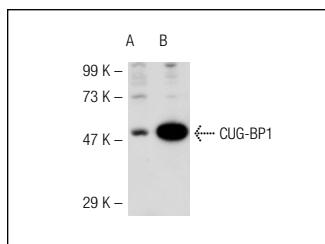
Molecular Weight of CUG-BP2: 54 kDa.

Positive Controls: CUG-BP1 (m2): 293T Lysate: sc-126681 or HeLa whole cell lysate: sc-2200.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 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 goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

DATA



CUG-BP1/2 (H-186): sc-48822. 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.

RESEARCH USE

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

MONOS
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Try **CUG-BP1/2 (B-1): sc-166095** or **CUG-BP1/2 (E-10): sc-166096**, our highly recommended monoclonal alternatives to CUG-BP1/2 (H-186).