

CUG-BP1/2 (E-10): sc-166096

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 upregulates 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 (CUG-BP1) 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 CUG-BP1 and alter protein levels and activity of CUG-BP1. *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: CUGBP1 (human) mapping to 11p11.2, CUGBP2 (human) mapping to 10p14; Cugbp1 (mouse) mapping to 2 E1, Cugbp2 (mouse) mapping to 2 A1.

SOURCE

CUG-BP1/2 (E-10) is a mouse monoclonal 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_{2b} kappa light chain 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 (E-10) is recommended for detection of CUG-BP1 and CUG-BP2 of mouse, rat and human origin by Western Blotting (starting dilution 1:100, 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).

Molecular Weight of CUG-BP1: 56 kDa.

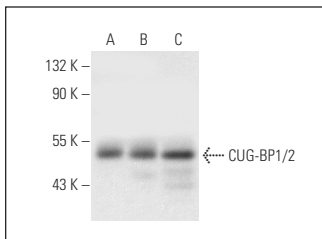
Molecular Weight of CUG-BP2: 54 kDa.

Positive Controls: HeLa whole cell lysate: sc-2200, Hep G2 cell lysate: sc-2227 or CUG-BP1 (m2): 293T Lysate: sc-126681.

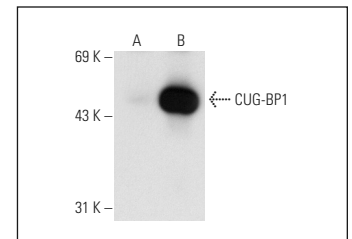
RECOMMENDED SUPPORT REAGENTS

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. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use m-IgG κ BP-FITC: sc-516140 or m-IgG κ 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



CUG-BP1/2 (E-10): sc-166096. Western blot analysis of CUG-BP1/2 expression in HeLa (A), Hep G2 (B) and HL-60 (C) whole cell lysates.



CUG-BP1/2 (E-10): sc-166096. 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.

PROTOCOLS

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



See **CUG-BP1 (3B1): sc-20003** for CUG-BP1 antibody conjugates, including AC, HRP, FITC, PE, and Alexa Fluor® 488, 546, 594, 647, 680 and 790.