

CUG-BP1 (3B1): sc-20003

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), hnRNP50. 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.

CHROMOSOMAL LOCATION

Genetic locus: CELF1 (human) mapping to 11p11.2; Celf1 (mouse) mapping to 2 E1.

SOURCE

CUG-BP1 (3B1) is a mouse monoclonal antibody raised against full length CUG-BP1 fusion protein of human origin.

PRODUCT

Each vial contains 200 μ g IgG₁ kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

CUG-BP1 (3B1) is available conjugated to agarose (sc-20003 AC), 500 μ g/0.25 ml agarose in 1 ml, for IP; to HRP (sc-20003 HRP), 200 μ g/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-20003 PE), fluorescein (sc-20003 FITC), Alexa Fluor® 488 (sc-20003 AF488), Alexa Fluor® 546 (sc-20003 AF546), Alexa Fluor® 594 (sc-20003 AF594) or Alexa Fluor® 647 (sc-20003 AF647), 200 μ g/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-20003 AF680) or Alexa Fluor® 790 (sc-20003 AF790), 200 μ g/ml, for Near-Infrared (NIR) WB, IF and FCM.

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RESEARCH USE

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

APPLICATIONS

CUG-BP1 (3B1) is recommended for detection of CUG-BP1 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 immunohistochemistry (including paraffin-embedded sections) (starting dilution 1:50, dilution range 1:50-1:500).

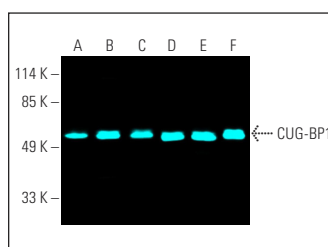
CUG-BP1 (3B1) is also recommended for detection of CUG-BP1 in additional species, including bovine and porcine.

Suitable for use as control antibody for CUG-BP1 siRNA (h): sc-38251, CUG-BP1 siRNA (m): sc-38252, CUG-BP1 shRNA Plasmid (h): sc-38251-SH, CUG-BP1 shRNA Plasmid (m): sc-38252-SH, CUG-BP1 shRNA (h) Lentiviral Particles: sc-38251-V and CUG-BP1 shRNA (m) Lentiviral Particles: sc-38252-V.

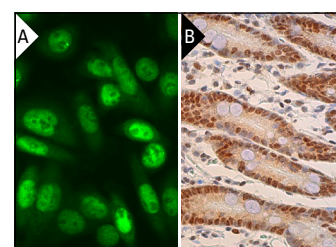
Molecular Weight of CUG-BP1: 56 kDa.

Positive Controls: HL-60 whole cell lysate: sc-2209, HeLa whole cell lysate: sc-2200 or NIH/3T3 whole cell lysate: sc-2210.

DATA



CUG-BP1 (3B1) Alexa Fluor® 647: sc-20003 AF647. Direct fluorescent western blot analysis of CUG-BP1 expression in HL-60 (A), HeLa (B), NIH/3T3 (C), Hep G2 (D) and c4 (E) whole cell lysates and HeLa nuclear extract (F). Blocked with UltraCruz® Blocking Reagent: sc-516214.



CUG-BP1 (3B1) Alexa Fluor® 488: sc-20003 AF488. Direct immunofluorescence staining of formalin-fixed SW480 cells showing nuclear localization. Blocked with UltraCruz® Blocking Reagent: sc-516214 (A). CUG-BP1 (3B1): sc-20003. Immunoperoxidase staining of formalin fixed, paraffin-embedded human duodenum tissue showing nuclear staining of glandular cells (B).

SELECT PRODUCT CITATIONS

1. Iakova, P., et al. 2004. Competition of CUG-BP1 and calreticulin for the regulation of p21 translation determines cell fate. *EMBO J.* 23: 406-417.
2. Liu, X., et al. 2017. Orthogonal ubiquitin transfer identifies ubiquitination substrates under differential control by the two ubiquitin activating enzymes. *Nat. Commun.* 8: 14286.
3. Dumbovic, G., et al. 2018. A novel long non-coding RNA from NBL2 pericentromeric macrosatellite forms a perinucleolar aggregate structure in colon cancer. *Nucleic Acids Res.* 46: 5504-5524.
4. Liu, Y., et al. 2019. A positive feedback regulation of Heme oxygenase 1 by CELF1 in cardiac myoblast cells. *Biochim. Biophys. Acta Gene Regul. Mech.* 1862: 209-218.

STORAGE

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