

CUG-BP2 (1H2): sc-47731

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.

CHROMOSOMAL LOCATION

Genetic locus: CELF2 (human) mapping to 10p14; Celf2 (mouse) mapping to 2 A1.

SOURCE

CUG-BP2 (1H2) is a mouse monoclonal antibody raised against amino acids 209-415 of CUG-BP2 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-BP2 (1H2) is available conjugated to agarose (sc-47731 AC), 500 μ g/0.25 ml agarose in 1 ml, for IP; to HRP (sc-47731 HRP), 200 μ g/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-47731 PE), fluorescein (sc-47731 FITC), Alexa Fluor® 488 (sc-47731 AF488), Alexa Fluor® 546 (sc-47731 AF546), Alexa Fluor® 594 (sc-47731 AF594) or Alexa Fluor® 647 (sc-47731 AF647), 200 μ g/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-47731 AF680) or Alexa Fluor® 790 (sc-47731 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

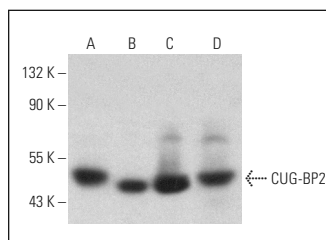
CUG-BP2 (1H2) is recommended for detection of CUG-BP2 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 μ g per 100-500 μ g of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and immunohistochemistry (including paraffin-embedded sections) (starting dilution 1:50, dilution range 1:50-1:500).

Suitable for use as control antibody for CUG-BP2 siRNA (h): sc-44554, CUG-BP2 siRNA (m): sc-44555, CUG-BP2 shRNA Plasmid (h): sc-44554-SH, CUG-BP2 shRNA Plasmid (m): sc-44555-SH, CUG-BP2 shRNA (h) Lentiviral Particles: sc-44554-V and CUG-BP2 shRNA (m) Lentiviral Particles: sc-44555-V.

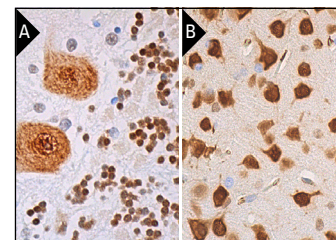
Molecular Weight of CUG-BP2: 54 kDa.

Positive Controls: EOC 20 whole cell lysate: sc-364187, Jurkat whole cell lysate: sc-2204 or CCRF-CEM cell lysate: sc-2225.

DATA



CUG-BP2 (1H2): sc-47731. Western blot analysis of CUG-BP2 expression in EOC 20 (A), Jurkat (B), CCRF-CEM (C) and SP2/0 (D) whole cell lysates.



CUG-BP2 (1H2): sc-47731. Immunoperoxidase staining of formalin fixed, paraffin-embedded human cerebellum tissue showing nuclear and cytoplasmic staining of Purkinje cells and nuclear staining of cells in granular layer and nuclear staining of cells in molecular layer (A). Immunoperoxidase staining of formalin fixed, paraffin-embedded mouse brain tissue showing nuclear and cytoplasmic staining of neuronal cells and glial cells and cytoplasmic staining of endothelial cells (B).

SELECT PRODUCT CITATIONS

1. Dujardin, G., et al. 2010. CELF proteins regulate CFTR pre-mRNA splicing: essential role of the divergent domain of ETR-3. *Nucleic Acids Res.* 38: 7273-7285.
2. Idris, M., et al. 2019. The MBNL/CELF splicing factors regulate cytosolic sulfotransferase 4A1 protein expression during cell differentiation. *Drug Metab. Dispos.* 47: 314-319.
3. Yoon, J.S.J., et al. 2020. Interleukin-10 control of pre-miR155 maturation involves CELF2. *PLoS ONE* 15: e0231639.
4. MacPherson, M.J., et al. 2021. Nucleocytoplasmic transport of the RNA-binding protein CELF2 regulates neural stem cell fates. *Cell Rep.* 35: 109226.

STORAGE

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