## SANTA CRUZ BIOTECHNOLOGY, INC.

# IGHMBP2 (N-14): sc-168143



### BACKGROUND

IGHMBP2 (immunoglobulin mu binding protein 2), is also known as HCSA, HMN6, CATF1, SMARD1 or SMUBP2, is a 993 amino acid nuclear and cytoplasmic protein that is ubiquitously expressed. Belonging to the DNA2/NAM7 helicase family, IGHMBP2 is a 5' to 3' helicase that unwinds RNA and DNA duplexes in an ATP-dependent reaction. IGHMBP2 also acts as a transcriptional regulator and is necessary for transcriptional activation of the flounder liver-type antifreeze protein gene. IGHMBP2 exists as a homooligomer and is part of the cytosolic ribonucleoprotein complex. Mutations in the gene encoding IGHMBP2 are suggested to lead to distal hereditary motor neuronopathy type 6 (HMN6), also known as spinal muscular atrophy distal autosomal recessive 1 (DSMA1) or spinal muscular atrophy with respiratory distress 1 (SMARD1). HMN6 is characterized by weakness and wasting of distal muscles of the anterior tibial and peroneal compartments of the legs and severe respiratory distress.

## REFERENCES

- 1. Fukita, Y., et al. 1993. The human S  $\mu$  bp-2, a DNA-binding protein specific to the single-stranded guanine-rich sequence related to the immunoglobulin mu chain switch region. J. Biol. Chem. 268: 17463-17470.
- Zhang, Q., et al. 1999. Smubp-2 represses the Epstein-Barr virus lytic switch promoter. Virology 255: 160-170.
- Grohmann, K., et al. 2001. Mutations in the gene encoding immunoglobulin μ-binding protein 2 cause spinal muscular atrophy with respiratory distress type 1. Nat. Genet. 29: 75-77.
- Grohmann, K., et al. 2003. Infantile spinal muscular atrophy with respiratory distress type 1 (SMARD1). Ann. Neurol. 54: 719-724.
- Liepinsh, E., et al. 2003. Solution structure of the R3H domain from human Smubp-2. J. Mol. Biol. 326: 217-223.
- Maystadt, I., et al. 2004. Allelic heterogeneity of SMARD1 at the IGHMBP2 locus. Hum. Mutat. 23: 525-526.
- Tachi, N., et al. 2005. A new mutation of IGHMBP2 gene in spinal muscular atrophy with respiratory distress type 1. Pediatr. Neurol. 32: 288-290.
- 8. de Planell-Saguer, M., et al. 2009. Biochemical and genetic evidence for a role of IGHMBP2 in the translational machinery. Hum. Mol. Genet. 18: 2115-2126.
- Guenther, U.P., et al. 2009. Clinical variability in distal spinal muscular atrophy type 1 (DSMA1): determination of steady-state IGHMBP2 protein levels in five patients with infantile and juvenile disease. J. Mol. Med. 87: 31-41.

#### CHROMOSOMAL LOCATION

Genetic locus: IGHMBP2 (human) mapping to 11q13.3; Ighmbp2 (mouse) mapping to 19 A.

#### SOURCE

IGHMBP2 (N-14) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the N-terminus of IGHMBP2 of human origin.

### PRODUCT

Each vial contains 200  $\mu g$  lgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-168143 P, (100  $\mu$ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

Available as TransCruz reagent for Gel Supershift and ChIP applications, sc-168143 X, 200  $\mu$ g/0.1 ml.

## **APPLICATIONS**

IGHMBP2 (N-14) is recommended for detection of IGHMBP2 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), 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).

IGHMBP2 (N-14) is also recommended for detection of IGHMBP2 in additional species, including canine.

Suitable for use as control antibody for IGHMBP2 siRNA (h): sc-96440, IGHMBP2 siRNA (m): sc-146184, IGHMBP2 shRNA Plasmid (h): sc-96440-SH, IGHMBP2 shRNA Plasmid (m): sc-146184-SH, IGHMBP2 shRNA (h) Lentiviral Particles: sc-96440-V and IGHMBP2 shRNA (m) Lentiviral Particles: sc-146184-V.

IGHMBP2 (N-14) X TransCruz antibody is recommended for Gel Supershift and ChIP applications.

Molecular Weight of IGHMBP2: 110 kDa.

#### **RECOMMENDED SECONDARY REAGENTS**

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker<sup>™</sup> compatible donkey anti-goat IgG-HRP: sc-2033 (dilution range: 1:2000-1:5000), Cruz Marker<sup>™</sup> Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunofluo-rescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz<sup>™</sup> Mounting Medium: sc-24941.

#### **STORAGE**

Store at 4° C, \*\*D0 NOT FREEZE\*\*. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

## **RESEARCH USE**

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

#### PROTOCOLS

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