SANTA CRUZ BIOTECHNOLOGY, INC.

# SMN (2B1): sc-32313

SANTA CRUZ BIOTECHNOLOGY

*The Power to Question*

## BACKGROUND

Spinal muscular atrophy (SMA) is an autosomal recessive neurodegenerative disease characterized by loss of motor neurons in the spinal cord. SMA is caused by deletion or loss-of-function mutations of SMN (survival of motor neuron) gene. SMN, also known as Gemin1, SMN1, SMNT and BCD541, exists as four isoforms produced by alternative splicing. SMN is oligomeric and forms a complex with Gemin2 (formerly SIP1), Gemin3 (a DEAD box RNA helicase), Gemin4, Gemin5 and Gemin6, as well as several spliceosomal snRNP proteins. The SMN complex plays an essential role in splicesomal snRNP assembly in the cytoplasm and is required for pre-mRNA splicing of the nucleus. The SMN complex is found in both the cytoplasm and the nucleus. The nuclear form is concentrated in subnuclear bodies called gems (gemini of the coiled bodies). Cytoplasmic SMN interacts with spliceosomal Sm proteins and facilitates their assembly onto U snRNAs, and nuclear SMN mediates recycling of pre-mRNA splicing factors. Nearly identical telomeric and centromeric forms of SMN encode the same protein; however, only mutations in the telomeric form are associated with the disease-state SMA. SMN is expresed in a wide variety of tissues including brain, kidney, liver, spinal cord and moderately in skeletal and cardiac muscle.

## REFERENCES


1. Coovert, D., et al. 1997. The survival motor neuron protein in spinal muscular atrophy. Hum. Mol. Genet. 6: 1205-1214.
2. Fischer, U., et al. 1997. The SMN-SIP1 complex has an essential role in spliceosomal snRNP biogenesis. Cell 90: 1023-1029.


## CHROMOSOMAL LOCATION

Genetic locus: SMN1 (human) mapping to 5q13.2; Smn1 (mouse) mapping to 13 D1.

## SOURCE

SMN (2B1) is a mouse monoclonal antibody raised against purified human recombinant His6 tagged-SMN protein.

## PRODUCT

Each vial contains 200 µg $IgG_1$ kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

SMN (2B1) is available conjugated to agarose (sc-32313 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-32313 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-32313 PE), fluorescein (sc-32313 FITC), Alexa Fluor® 488 (sc-32313 AF488), Alexa Fluor® 546 (sc-32313 AF546), Alexa Fluor® 594 (sc-32313 AF594) or Alexa Fluor® 647 (sc-32313 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-32313 AF680) or Alexa Fluor® 790 (sc-32313 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.



## STORAGE

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

## APPLICATIONS

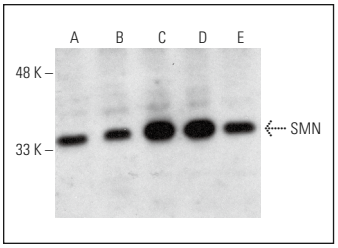
SMN (2B1) is recommended for detection of SMN of mouse, rat, human and *Xenopus laevis* origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution 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 SMN siRNA (h): sc-36510, SMN siRNA (m): sc-36511, SMN shRNA Plasmid (h): sc-36510-SH, SMN shRNA Plasmid (m): sc-36511-SH, SMN shRNA (h) Lentiviral Particles: sc-36510-V and SMN shRNA (m) Lentiviral Particles: sc-36511-V.

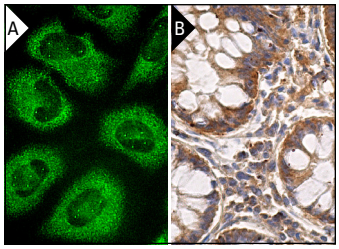
Molecular Weight of SMN: 39 kDa.

Positive Controls: HeLa whole cell lysate: sc-2200, HeLa nuclear extract: sc-2120 or Jurkat nuclear extract: sc-2132.

## DATA




SMN (2B1) HRP: sc-32313 HRP. Direct western blot analysis of SMN expression in HeLa (**A**), Jurkat (**B**) and 293 (**C**) whole cell lysates and HeLa (**D**) and Jurkat (**E**) nuclear extracts.




SMN (2B1): sc-32313. Immunofluorescence staining of methanol-fixed HeLa cells showing cytoplasmic and cajal bodies localization (**A**). Immunoperoxidase staining of formalin fixed, paraffin-embedded human colon tissue showing cytoplasmic staining of glandular cells (**B**).

## SELECT PRODUCT CITATIONS


1. Pellizzoni, L., et al. 2002. Purification of native survival of motor neurons complexes and identification of Gemin6 as a novel component. J. Biol. Chem. 277: 7540-7545.
2. Doktor, T.K., et al. 2016. RNA-sequencing of a mouse-model of spinal muscular atrophy reveals tissue-wide changes in splicing of U12-dependent introns. Nucleic Acids Res. 45: 395-416.
3. Zhang, Q.J., et al. 2017. Application of urine cells in drug intervention for spinal muscular atrophy. Exp. Ther. Med. 14: 1993-1998.
4. Wang, H., et al. 2018. CRISPR-mediated programmable 3D genome positioning and nuclear organization. Cell 175: 1405-1417.e13.
5. Ellwanger, K., et al. 2019. XIAP controls RIPK2 signaling by preventing its deposition in speck-like structures. Life Sci. Alliance 2: e201900346.
6. Roithová, A., et al. 2020. DIS3L2 and LSm proteins are involved in the surveillance of Sm ring-deficient snRNAs. Nucleic Acids Res. 48: 6184-6197.


## RESEARCH USE

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