SANTA CRUZ BIOTECHNOLOGY, INC.

SMN (H-195): sc-15320

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 (for 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.

CHROMOSOMAL LOCATION

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

SOURCE

SMN (H-195) is a rabbit polyclonal antibody raised against amino acids 1-195 mapping at the N-terminus of SMN of human origin.

PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

SMN (H-195) is recommended for detection of SMN of mouse, rat and human 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).

SMN (H-195) is also recommended for detection of SMN in additional species, including porcine.

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.

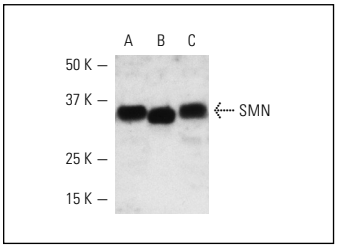
Molecular Weight of SMN: 39 kDa.

Positive Controls: SK-BR-3 nuclear extract: sc-2134, K-562 nuclear extract: sc-2130 or HeLa nuclear extract: sc-2120.

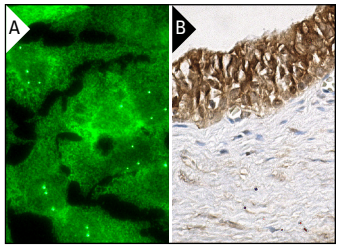
STORAGE

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

DATA



SMN (H-195): sc-15320. Western blot analysis of SMN expression in HeLa (**A**), SK-BR-3 (**B**) and K-562 (**C**) nuclear extracts.



SMN (H-195): sc-15320. Immunofluorescence staining of formalin-fixed Hep G2 cells showing cytoplasmic and nuclear localization (**A**). Immunoperoxidase staining of formalin fixed, paraffin-embedded human testis tissue showing cytoplasmic and nuclear staining of cells in seminiferous ducts (**B**).

SELECT PRODUCT CITATIONS

1. Hua, Y., et al. 2004. Survival motor neuron protein facilitates assembly of stress granules. FEBS Lett. 572: 69-74.
2. Bilinski, S.M., et al. 2004. Sm proteins, the constituents of the spliceosome, are components of nuage and mitochondrial cement in *Xenopus* oocytes. Exp. Cell Res. 299: 171-178.
3. Zelazowska, M. and Jaglarz, M.K., 2004. Oogenesis in phthirapterans *(Insecta: Phthiraptera).* I. Morphological and histochemical characterization of the oocyte nucleus and its inclusions. Arthropod Struct. Dev. 33: 161-172.
4. Gao, X., et al. 2012. Tudor staphylococcal nuclease (Tudor-SN) participates in small ribonucleoprotein (snRNP) assembly via interacting with symmetrically dimethylated Sm proteins. J. Biol. Chem. 287: 18130-18141.
5. Tapia, O., et al. 2012. Reorganization of Cajal bodies and nucleolar targeting of coilin in motor neurons of type I spinal muscular atrophy. Histochem. Cell Biol. E-published.
6. Tsuiji, H., et al. 2013. Spliceosome integrity is defective in the motor neuron diseases ALS and SMA. EMBO Mol. Med. 5: 221-234.
7. Rafałowska, J., et al. 2014. Diverse expression of selected SMN complex proteins in humans with sporadic amyotrophic lateral sclerosis and in a transgenic rat model of familial form of the disease. PLoS ONE 9: e104614.

RESEARCH USE

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