SANTA CRUZ BIOTECHNOLOGY, INC.

SMN (11F3): sc-81554

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.
3. Monani, U., et al. 1999. A single nucleotide difference that alters splicing patterns distinguishes the SMA gene SMN1 from the copy gene SMN2. Hum. Mol. Genet. 8: 1177-1183.
4. Meister, G., et al. 2000. Characterization of a nuclear 20S complex containing the survival of motor neurons (SMN) protein and a specific subset of spliceosomal Sm proteins. Hum. Mol. Genet. 9: 1977-1986.
5. Mourelatos, Z., et al. 2001. SMN interacts with a novel family of hnRNP and spliceosomal proteins. EMBO J. 20: 5443-5452.
6. Online Mendelian Inheritance in Man, OMIM™. 2002. Johns Hopkins University, Baltimore, MD. MIM Number: 600354. World Wide Web URL: http://www.ncbi.nlm.nih.gov/omim/
7. LocusLink Report (LocusID: 6606). http://www.ncbi.nlm.nih.gov/LocusLink/

CHROMOSOMAL LOCATION

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

SOURCE

SMN (11F3) is a mouse monoclonal antibody raised against full length recombinant SMN of human origin, with epitope mapping to amino acids 28-91.

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.

APPLICATIONS

SMN (11F3) 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)] and immunofluorescence (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.

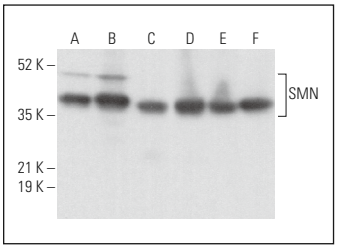
Molecular Weight of SMN: 39 kDa.

Positive Controls: SMN (h2): 293T Lysate: sc-113432, HeLa whole cell lysate: sc-2200 or MDA-MB-231 cell lysate: sc-2232.

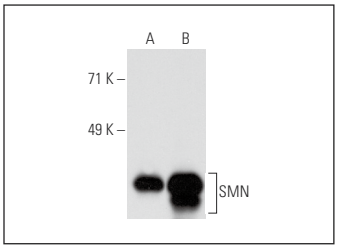
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

DATA



SMN (11F3): sc-81554. Western blot analysis of SMN expression in HeLa (**A**), MDA-MB-231 (**B**), BT-20 (**C**), Neuro-2A (**D**), F9 (**E**) and AT3B-1 (**F**) whole cell lysates.



SMN (11F3): sc-81554. Western blot analysis of SMN expression in non-transfected: sc-117752 (**A**) and human SMN transfected: sc-113432 (**B**) 293T whole cell lysates.

STORAGE

Store at 4° C, **DO 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.