SANTA CRUZ BIOTECHNOLOGY, INC.

Gemin6 (20H8): sc-130667

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 in the SMN (survival of motor neuron) gene. Gemin6, the protein product of human chromosome 2p22.1, associates directly with SMN and is a part of the SMN complex containing Gemin2, Gemin3, Gemin4 and Gemin5 as well as several spliceosomal snRNP proteins. The SMN complex plays an essential role in spliceosomal 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).

REFERENCES

1. Coovert, D., Le, T., McAndrew, P., Strasswimmer, J., Crawford, T., Mendell, J., Coulson, S., Androphy, E., Prior, T. and Burghes, A. 1997. The survival motor neuron protein in spinal muscular atrophy. Hum. Mol. Genet. 6: 1205-1214.
2. Fischer, U., Liu, Q. and Dreyfuss, G. 1997. The SMN-SIP1 complex has an essential role in spliceosomal snRNP biogenesis. Cell 90: 1023-1029.
3. Monani, U., Lorson, C., Parsons, D., Prior, T., Androphy, E., Borghes, A. and McPherson, J. 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., Buhler, D., Lafferbauer, B., Zobawa, M., Lottspeich, F. and Fisher, U. 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., Abel, L., Yong, J., Kataoka, N. and Dreyfuss, G. 2001. SMN interacts with a novel family of hnRNP and spliceosomal proteins. EMBO J. 20: 5443-5452.
6. Pellizzoni, L., Baccon, J., Rappsilber, J., Mann, M. and Dreyfuss, G. 2002. Purification of native survival of motor neuron complexes and identification of Gemin6 as a novel component. J. Biol. Chem. 277: 7540-7545.

CHROMOSOMAL LOCATION

Genetic locus: GEMIN6 (human) mapping to 2p22.1.

SOURCE

Gemin6 (20H8) is a mouse monoclonal antibody raised against recombinant Gemin6 of human origin.

PRODUCT

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

STORAGE

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

APPLICATIONS

Gemin6 (20H8) is recommended for detection of Gemin6 of 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 Gemin6 siRNA (h): sc-42132, Gemin6 shRNA Plasmid (h): sc-42132-SH and Gemin6 shRNA (h) Lentiviral Particles: sc-42132-V.

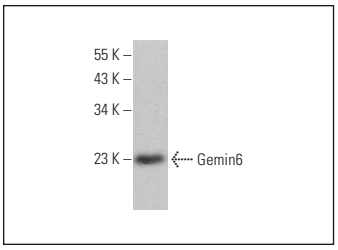
Molecular Weight of Gemin6: 19 kDa.

Positive Controls: Jurkat whole cell lysate: sc-2204

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, TBS Blotto A Blocking Reagent: sc-2333 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



Gemin6 (20H8): sc-130667. Western blot analysis of Gemin6 expression in Jurkat whole cell lysate. Detection reagent used: m-IgGκ BP-HRP: sc-516102.

SELECT PRODUCT CITATIONS

1. Yamazaki, T., Chen, S., Yu, Y., Yan, B., Haertlein, T.C., Carrasco, M.A., Tapia, J.C., Zhai, B., Das, R., Lalancette-Hebert, M., Sharma, A., Chandran, S., Sullivan, G., Nishimura, A.L., Shaw, C.E., Gygi, S.P., et al. 2012. FUS-SMN protein interactions link the motor neuron diseases ALS and SMA. Cell Rep. 2: 799-806.
2. Yu, Y., Chi, B., Xia, W., Gangopadhyay, J., Yamazaki, T., Winkelbauer-Hurt, M.E., Yin, S., Eliasse, Y., Adams, E., Shaw, C.E. and Reed, R. 2015. U1 snRNP is mislocalized in ALS patient fibroblasts bearing NLS mutations in FUS and is required for motor neuron outgrowth in zebrafish. Nucleic Acids Res. 43: 3208-3218.

RESEARCH USE

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