

SMN1 Antibody

Purified Mouse Monoclonal Antibody

Catalog # AO1572a

Product Information

Application	WB, IHC, ICC, E
Primary Accession	Q16637
Reactivity	Human, Monkey
Host	Mouse
Clonality	Monoclonal
Clone Names	5H1
Isotype	IgG1
Calculated MW	31849
Description	This gene is part of a 500 kb inverted duplication on chromosome 5q13. This duplicated region contains at least four genes and repetitive elements which make it prone to rearrangements and deletions. The repetitiveness and complexity of the sequence have also caused difficulty in determining the organization of this genomic region. The telomeric and centromeric copies of this gene are nearly identical and encode the same protein.
Immunogen	Purified recombinant fragment of human SMN1 expressed in E. Coli.
Formulation	Ascitic fluid containing 0.03% sodium azide.

Additional Information

Gene ID	6606;6607
Other Names	Survival motor neuron protein, Component of gems 1, Gemin-1, SMN1, SMN, SMNT
Dilution	WB~~1/500 - 1/2000 IHC~~1/200 - 1/1000 ICC~~N/A E~~1/10000
Storage	Maintain refrigerated at 2-8°C for up to 6 months. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.
Precautions	SMN1 Antibody is for research use only and not for use in diagnostic or therapeutic procedures.

Protein Information

Name	SMN1
Synonyms	SMN, SMNT

Function	<p>The SMN complex catalyzes the assembly of small nuclear ribonucleoproteins (snRNPs), the building blocks of the spliceosome, and thereby plays an important role in the splicing of cellular pre- mRNAs (PubMed:18984161, PubMed:9845364). Most spliceosomal snRNPs contain a common set of Sm proteins SNRNPB, SNRPD1, SNRPD2, SNRPD3, SNRPE, SNRPF and SNRPG that assemble in a heptameric protein ring on the Sm site of the small nuclear RNA to form the core snRNP (Sm core) (PubMed:18984161). In the cytosol, the Sm proteins SNRPD1, SNRPD2, SNRPE, SNRPF and SNRPG are trapped in an inactive 6S pICln-Sm complex by the chaperone CLNS1A that controls the assembly of the core snRNP (PubMed:18984161). To assemble core snRNPs, the SMN complex accepts the trapped 5Sm proteins from CLNS1A forming an intermediate (PubMed:18984161). Within the SMN complex, SMN1 acts as a structural backbone and together with GEMIN2 it gathers the Sm complex subunits (PubMed:17178713, PubMed:21816274, PubMed:22101937). Binding of snRNA inside 5Sm ultimately triggers eviction of the SMN complex, thereby allowing binding of SNRPD3 and SNRNPB to complete assembly of the core snRNP (PubMed:31799625). Ensures the correct splicing of U12 intron- containing genes that may be important for normal motor and proprioceptive neurons development (PubMed:23063131). Also required for resolving RNA-DNA hybrids created by RNA polymerase II, that form R- loop in transcription terminal regions, an important step in proper transcription termination (PubMed:26700805). May also play a role in the metabolism of small nucleolar ribonucleoprotein (snoRNPs).</p>
Cellular Location	<p>Nucleus, gem. Nucleus, Cajal body. Cytoplasm. Cytoplasmic granule. Perikaryon. Cell projection, neuron projection. Cell projection, axon {ECO:0000250 UniProtKB:P97801}. Cytoplasm, myofibril, sarcomere, Z line {ECO:0000250 UniProtKB:P97801}. Note=Colocalizes with actin and at the Z-line of skeletal muscle (By similarity). Under stress conditions colocalizes with RPP20/POP7 in punctuated cytoplasmic granules (PubMed:14715275). Colocalized and redistributed with ZPR1 from the cytoplasm to nuclear gems (Gemini of coiled bodies) and Cajal bodies (PubMed:11283611). Colocalizes with FMR1 in cytoplasmic granules in the soma and neurite cell processes (PubMed:18093976) {ECO:0000250 UniProtKB:P97801, ECO:0000269 PubMed:11283611, ECO:0000269 PubMed:14715275, ECO:0000269 PubMed:18093976}</p>
Tissue Location	<p>Expressed in a wide variety of tissues. Expressed at high levels in brain, kidney and liver, moderate levels in skeletal and cardiac muscle, and low levels in fibroblasts and lymphocytes. Also seen at high levels in spinal cord. Present in osteoclasts and mononuclear cells (at protein level).</p>

References

1. J Med Genet. 2009 Sep;46(9):641-4. 2. RNA. 2009 Apr;15(4):515-23.

Images

Figure 1: Western blot analysis using SMN1 mouse mAb against RAJI (1), Cos7 (2), Jurkat (3), K562 (4), Hela (5) and HepG2 (6) cell lysate.

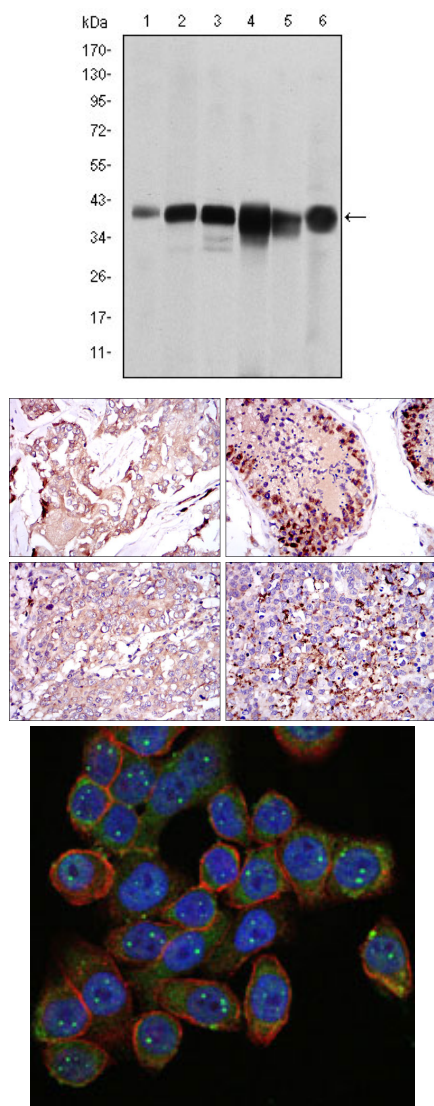


Figure 2: Immunohistochemical analysis of paraffin-embedded breast cancer tissues (left) and testis tissues (right) using SMN1 mouse mAb with DAB staining.

Figure 3: Immunohistochemical analysis of paraffin-embedded stomach cancer tissues (left) and brain tumor (right) using SMN1 mouse mAb with DAB staining.

Figure 4: Immunofluorescence analysis of HepG2 cells using SMN1 mouse mAb (green). Blue: DRAQ5 fluorescent DNA dye. Red: Actin filaments have been labeled with Alexa Fluor-555 phalloidin.

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