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SMN1 Antibody (C-term) Blocking Peptide

Synthetic peptide
Catalog # BP18621b

Specification

SMN1 Antibody (C-term) Blocking Peptide - Product Information

Primary Accession Q16637

SMN1 Antibody (C-term) Blocking Peptide - Additional Information

Gene ID 6606;6607

Other Names
Survival motor neuron protein, Component of gems 1, Gemin-1, SMN1, SMN, SMNT

Format
Peptides are lyophilized in a solid powder format. Peptides can be reconstituted in solution using the appropriate buffer as needed.

Storage
Maintain refrigerated at 2-8°C for up to 6 months. For long term storage store at -20°C.

Precautions
This product is for research use only. Not for use in diagnostic or therapeutic procedures.

SMN1 Antibody (C-term) Blocking Peptide - Protein Information

Name SMN1

Synonyms SMN, SMNT

Function
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:9845364, PubMed:18984161). Most spliceosomal snRNPs contain a common set of Sm proteins SNRPB, 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:<a href="http://www.uniprot.org/citations/21816274"

target="_blank">21816274, PubMed:22101937, PubMed:17178713). Binding of snRNA inside 5Sm ultimately triggers eviction of the SMN complex, thereby allowing binding of SNRPD3 and SNRPB 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).

Cellular Location
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}

Tissue Location
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).

SMN1 Antibody (C-term) Blocking Peptide - Protocols

Provided below are standard protocols that you may find useful for product applications.

- Blocking Peptides

SMN1 Antibody (C-term) Blocking Peptide - Images

SMN1 Antibody (C-term) Blocking Peptide - Background

This gene is part of a 500 kb inverted duplication onchromosome 5q13. This duplicated region contains at least fourgenes and repetitive elements which make it prone to rearrangementsand deletions. The repetitiveness and complexity of the sequencehave also caused difficulty in determining the organization of thisgenomic region. The telomeric and centromeric copies of this geneare nearly identical and encode the same protein. However,mutations in this gene, the telomeric copy, are associated withspinal muscular atrophy; mutations in the centromeric copy do notlead to disease. The centromeric copy may be a modifier of diseasecaused by mutation in the telomeric copy. The critical sequencedifference between the two genes is a single nucleotide in exon 7,which is thought to be an exon splice enhancer. Note that the nineexons of both the telomeric and centromeric copies are designatedhistorically as exon 1, 2a, 2b, and 3-8. It is thought that geneconversion events may involve the two genes, leading to varyingcopy numbers of each gene. The protein encoded by this genelocalizes to both the cytoplasm and the nucleus. Within thenucleus, the protein localizes to subnuclear bodies called gemswhich are found near coiled bodies containing high concentrationsof small ribonucleoproteins (snRNPs). This protein formsheteromeric complexes with proteins such as SIP1 and GEMIN4, andalso interacts with several proteins known to be involved in thebiogenesis of snRNPs, such as hnRNP U protein and the

smallnucleolar RNA binding protein. Two transcript variants encodingdistinct isoforms have been described.

SMN1 Antibody (C-term) Blocking Peptide - References

Todd, A.G., et al. J. Mol. Biol. 401(5):681-689(2010)Sheng-Yuan, Z., et al. Eur. J. Hum. Genet. 18(9):978-984(2010)Liu, W.L., et al. Zhongguo Dang Dai Er Ke Za Zhi 12(7):539-543(2010)Wang, C.C., et al. Anal Bioanal Chem 397(6):2375-2383(2010)Bebee, T.W., et al. Front. Biosci. 15, 1191-1204 (2010) :