

# Product datasheet

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# ARG42778 anti-SMN / Gemin1 antibody

Package: 100 μl Store at: -20°C

#### Summary

Product Description Rabbit Polyclonal antibody recognizes SMN / Gemin1

Tested Reactivity Hu, Ms, Rat

Tested Application IHC-P, IP, WB

Host Rabbit

**Clonality** Polyclonal

Isotype IgG

Target Name SMN / Gemin1

Species Human

Immunogen Synthetic peptide derived from Human SMN / Gemin1.

Conjugation Un-conjugated

Alternate Names SMA3; SMA2; SMA1; Component of gems 1; SMNT; SMA4; GEMIN1; Survival motor neuron protein;

BCD541; SMN; SMA@; T-BCD541; TDRD16A; SMA; Gemin-1

### **Application Instructions**

Application table	Application	Dilution
	IHC-P	1:50 - 1:200
	IP	1:50
	WB	1:500 - 1:2000
Application Note	* The dilutions indicate recommended starting dilutions and the optimal dilutions or concentrations should be determined by the scientist.	
Positive Control	HepG2	
Observed Size	~ 36 kDa	

#### **Properties**

Form Liquid

Purification Affinity purified.

Buffer PBS (pH 7.4), 150 mM NaCl, 0.02% Sodium azide and 50% Glycerol.

Preservative 0.02% Sodium azide

Stabilizer 50% Glycerol

Storage instruction For continuous use, store undiluted antibody at 2-8°C for up to a week. For long-term storage, aliquot

and store at -20°C. Storage in frost free freezers is not recommended. Avoid repeated freeze/thaw cycles. Suggest spin the vial prior to opening. The antibody solution should be gently mixed before use.

#### Bioinformation

Gene Symbol

SMN1

Gene Full Name

survival of motor neuron 1, telomeric

Background

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. However, mutations in this gene, the telomeric copy, are associated with spinal muscular atrophy; mutations in the centromeric copy do not lead to disease. The centromeric copy may be a modifier of disease caused by mutation in the telomeric copy. The critical sequence difference between the two genes is a single nucleotide in exon 7, which is thought to be an exon splice enhancer. Note that the nine exons of both the telomeric and centromeric copies are designated historically as exon 1, 2a, 2b, and 3-8. It is thought that gene conversion events may involve the two genes, leading to varying copy numbers of each gene. The protein encoded by this gene localizes to both the cytoplasm and the nucleus. Within the nucleus, the protein localizes to subnuclear bodies called gems which are found near coiled bodies containing high concentrations of small ribonucleoproteins (snRNPs). This protein forms heteromeric complexes with proteins such as SIP1 and GEMIN4, and also interacts with several proteins known to be involved in the biogenesis of snRNPs, such as hnRNP U protein and the small nucleolar RNA binding protein. Multiple transcript variants encoding distinct isoforms have been described. [provided by RefSeq, Jul 2014]

Function

The SMN complex plays a catalyst role in the assembly of small nuclear ribonucleoproteins (snRNPs), the building blocks of the spliceosome. Thereby, plays an important role in the splicing of cellular premRNAs. 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. In the cytosol, the Sm proteins SNRPD1, SNRPD2, SNRPE, SNRPF and SNRPG are trapped in an inactive 6S plCln-Sm complex by the chaperone CLNS1A that controls the assembly of the core snRNP. Dissociation by the SMN complex of CLNS1A from the trapped Sm proteins and their transfer to an SMN-Sm complex triggers the assembly of core snRNPs and their transport to the nucleus. Ensures the correct splicing of U12 intron-containing genes that may be important for normal motor and proprioceptive neurons development. 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. May also play a role in the metabolism of small nucleolar ribonucleoprotein (snoRNPs). [UniProt]

Calculated Mw

32 kDa

Cellular Localization

Nucleus, gem. Nucleus, Cajal body. Cytoplasm. Cytoplasmic granule. Perikaryon. Cell projection. Cytoplasm, myofibril, sarcomere, Z line. Note=Colocalizes with actin and at the Z-line of skeletal muscle. Under stress conditions colocalizes with RPP20/POP7 in punctuated cytoplasmic granules. Colocalized and redistributed with ZPR1 from the cytoplasm to nuclear gems and Cajal bodies. Colocalizes with FMR1 in cytoplasmic granules in the soma and neurite cell processes. [UniProt]



## ARG42778 anti-SMN / Gemin1 antibody WB image

Western blot: HepG2 cell lysate stained with ARG42778 anti-SMN /  ${\tt Gemin1}$  antibody.