SMN1 Antibody – Rabbit Polyclonal

PRODUCT OVERVIEW

Product Name	SMN1, Polyclonal Antibody
Full Product Name	SMN1, CT (SMN1, SMN, SMNT, Survival motor neuron protein, Component of gems 1, Gemin-1) (MaxLight 650)
Product Gene Name	anti-SMN1 antibody
Product Synonym Names	SMN1; SMN; SMNT; Survival motor neuron protein; Component of gems 1; Gemin-1
Clonality	Polyclonal
Isotype	IgG
Host	Rabbit
Species Reactivity	Human
Form/Format	Supplied as a liquid in PBS, pH 7.2. No preservative added. Labeled with MaxLight650.
Purity/Purification	Purified by Protein A Affinity Chromatography.
Specificity	Recognizes human SMN1.
Preparation and Storage	Store product at 4 degree C in the dark. DO NOT FREEZE! Stable at 4 degree C for 12 months after receipt as an undiluted liquid. Dilute required amount only prior to immediate use. Further dilutions can be made in assay buffer. Caution: MaxLight650 conjugates are sensitive to light. For maximum recovery of product, centrifuge the original vial prior to removing the cap.
Molecular Weight	27,135 Da
OMIM	253300
3D Structure	ModBase 3D Structure for Q16637
Immunogen	KLH-conjugated synthetic peptide mapping to a fragment of residues within amino acids 262-288 in the C-terminal region of human SMN1, UniProt Accession #Q16637. Species sequence homology: monkey
Conjugate	MaxLight650
Note	Preservative Free
Product Categories/Family for anti-SMN1 antibody	Antibodies; Proteins
Applications Tested/Suitable for anti-SMN1 antibody	Western Blot (WB), FLISA
Application Notes for anti-SMN1 antibody	WB: 1:1000 Applications are based on unconjugated antibody.
Related Product Information for anti-SMN1 antibody	MBS6348976 is an antibody targeting SMN1. Also known as SMN1, CT (SMN1, SMN, SMNT, Survival motor neuron protein, Component of gems 1, Gemin-1) (MaxLight 650). Reactive in Human. Host: Rabbit.
Product References and Citations for anti-SMN1 antibody	1. Todd, A.G., et al. J. Mol. Biol. 401(5):681-689(2010). 2. Sheng-Yuan, Z., et al. Eur. J. Hum. Genet. 18(9):978-984(2010). 3. Liu, W.L., et al. Zhongguo Dang Dai Er Ke Za Zhi 12(7):539-543(2010). 4. Wang, C.C., et al. Anal Bioanal Chem 397(6):2375-2383(2010). 5. Bebee, T.W., et al. Front. Biosci. 15, 1191-1204 (2010).
Other Notes	Small volumes of anti-SMN1 antibody vial(s) may occasionally become entrapped in the seal of the product vial during shipment and storage. If necessary, briefly centrifuge the vial on a tabletop centrifuge to dislodge any liquid in the container`s cap. Certain products may require to ship with dry ice and additional dry ice fee may apply.

ADDITIONAL DETAILS

NCBI/Uniprot data below describe general gene information for SMN1. It may not necessarily be applicable to this product.

NCBI GI #	4507091
NCBI GeneID	6606
NCBI Accession #	NP_000335.1
UniProt Primary Accession #	Q16637
UniProt Secondary Accession #	Q13119; Q549U5; Q96J51; A8K0V4
UniProt Related Accession #	Q16637
GenBank Nucleotide #	NM_000344.3
NCBI Official Full Name	survival motor neuron protein isoform d
NCBI Official Synonym Full Names	survival of motor neuron 1, telomeric
NCBI Official Symbol	SMN1
NCBI Official Synonym Symbols	SMA; SMN; SMA1; SMA2; SMA3; SMA4; SMA@; SMNT; BCD541; GEMIN1; TDRD16A; T-BCD541
UniProt Protein Name	Survival motor neuron protein
UniProt Synonym Protein Names	Component of gems 1; Gemin-1
Protein Family	Survival motor neuron-like protein
UniProt Gene Name	SMN1
UniProt Synonym Gene Names	SMN; SMNT
UniProt Entry Name	SMN_HUMAN
NCBI Protein Information	survival motor neuron protein
NCBI Summary for SMN1	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]

UniProt Comments for SMN1	SMN: The SMN complex plays an essential role in spliceosomal snRNP assembly in the cytoplasm and is required for pre-mRNA splicing in the nucleus. It may also play a role in the metabolism of snoRNPs. Defects in SMN1 are the cause of spinal muscular atrophy autosomal recessive type 1 (SMA1). Spinal muscular atrophy refers to a group of neuromuscular disorders characterized by degeneration of the anterior horn cells of the spinal cord, leading to symmetrical muscle weakness and atrophy. Autosomal recessive forms are classified according to the age of onset, the maximum muscular activity achieved, and survivorship. The severity of the disease is mainly determined by the copy number of SMN2, a copy gene which predominantly produces exon 7-skipped transcripts and only low amount of full-length transcripts that encode for a protein identical to SMN1. Only about 4% of SMA patients bear one SMN1 copy with an intragenic mutation. SMA1 is a severe form, with onset before 6 months of age. SMA1 patients never achieve the ability to sit. Defects in SMN1 are the cause of spinal muscular atrophy autosomal recessive type 2 (SMA2). SMA2 is an autosomal recessive spinal muscular atrophy of intermediate severity, with onset between 6 and 18 months. Patients do not reach the motor milestone of standing, and survive into adulthood. Defects in SMN1 are the cause of spinal muscular atrophy autosomal recessive type 3 (SMA3). SMA3 is an autosomal recessive spinal muscular atrophy with onset after 18 months. SMA3 patients develop ability to stand and walk and survive into adulthood. Defects in SMN1 are the cause of spinal muscular atrophy autosomal recessive type 4 (SMA4). SMA4 is an autosomal recessive spinal muscular atrophy characterized by symmetric proximal muscle weakness with onset in adulthood and slow disease progression. SMA4 patients can stand and walk. Belongs to the SMN family. 4 isoforms of the human protein are produced by alternative splicing. Protein type: RNA-binding; RNA processing Chromosomal Location of Human Ortholog: 5q13.2 Cellular Component: nucleoplasm; spliceosome; Cajal body; cytoplasm; SMN complex; nucleus; Z disc; cytosol Molecular Function: identical protein binding; protein binding; RNA binding Biological Process: nervous system development; spliceosomal snRNP biogenesis; spliceosome assembly; gene expression Disease: Spinal Muscular Atrophy, Type Ii; Spinal Muscular Atrophy, Type Iii; Spinal Muscular Atrophy, Type Iv; Spinal Muscular Atrophy, Type I
Research Articles on SMN1	**1.** The diverse a-SMN vs FL-SMN C-terminus may dictate different protein interactions and complex formation explaining the different localization and role in the neuronal compartment, and the lower expression and stability of a-SMN.
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