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Rabbit Anti-SMN1 antibody

SL4628R

Product Name:	SMN1
Chinese Name:	运动神经元生存蛋白1抗体
Alias:	Component of gems 1; Component of gems 2; Gemin 1; Gemin-1; SMA; SMA1; SMA3; SMN; SMN_HUMAN; SMN1; SMN2; SMNC; SMNT; Survival motor neuron protein; survival of motor neuron 1, telomeric; survival of motor neuron 2, centromeric.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800Flow-Cyt=1ug/TestIF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	32kDa
Cellular localization:	The nucleuscytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human SMN1:51-150/294
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
PubMed:	PubMed
Product Detail:	SMN1 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. While mutations in the telomeric copy are associated with spinal muscular atrophy, mutations in this gene, the centromeric copy, do not lead to disease. This gene 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 full length 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. Four transcript variants encoding distinct isoforms have been described. **Function:** 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. **Subunit:** Component of an import snRNP complex composed of KPNB1, RNUT1, SMN1 and ZNF259. Part of the core SMN complex that contains SMN1, GEMIN2/SIP1, DDX20/GEMIN3, GEMIN4, GEMIN5, GEMIN6, GEMIN7, GEMIN8 and STRAP/UNRIP. Interacts with DDX20, FBL, NOLA1, RNUT1, SYNCRIP and with several spliceosomal snRNP core Sm proteins, including SNRPB, SNRPD1, SNRPD2, SNRPD3, SNRPE and ILF3. Interacts with OSTF1, LSM10 and LSM11. **Subcellular Location:** Cytoplasm. Nucleus, gem. Note=Localized in subnuclear structures next to coiled bodies, called Gemini of Cajal bodies (Gems). **Tissue Specificity:** 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). **DISEASE:** Spinal muscular atrophy 1 (SMA1) [MIM:253300]: A form of spinal muscular atrophy, a group of neuromuscular disorder 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. Note=The disease is caused by mutations affecting the gene represented in this entry.
Spinal muscular atrophy 2 (SMA2) [MIM:253550]: An autosomal recessive form of spinal muscular atrophy, a neuromuscular disorder characterized by degeneration of the anterior horn cells of the spinal cord, leading to symmetrical muscle weakness and atrophy. It has intermediate severity, with onset between 6 and 18 months. Patients do not reach the motor milestone of standing, and survive into adulthood. Note=The disease is caused by mutations affecting the gene represented in this entry.
Spinal muscular atrophy 3 (SMA3) [MIM:253400]: An autosomal recessive form of spinal muscular atrophy, a neuromuscular disorder characterized by degeneration of the anterior horn cells of the spinal cord, leading to symmetrical muscle weakness and atrophy. Onset is after 18 months. Patients develop ability to stand and walk and survive into adulthood. Note=The disease is caused by mutations affecting the gene represented in this entry.
Spinal muscular atrophy 4 (SMA4) [MIM:271150]: An autosomal recessive form of spinal muscular atrophy, a neuromuscular disorder characterized by degeneration of the anterior horn cells of the spinal cord, leading to symmetrical muscle weakness and atrophy. Onset is in adulthood, disease progression is slow, and patients can stand and walk. Note=The disease is caused by mutations affecting the gene represented in this entry.

Similarity:
Belongs to the SMN family.
Contains 1 Tudor domain.

SWISS:
O14893

Gene ID:
6606

Database links:

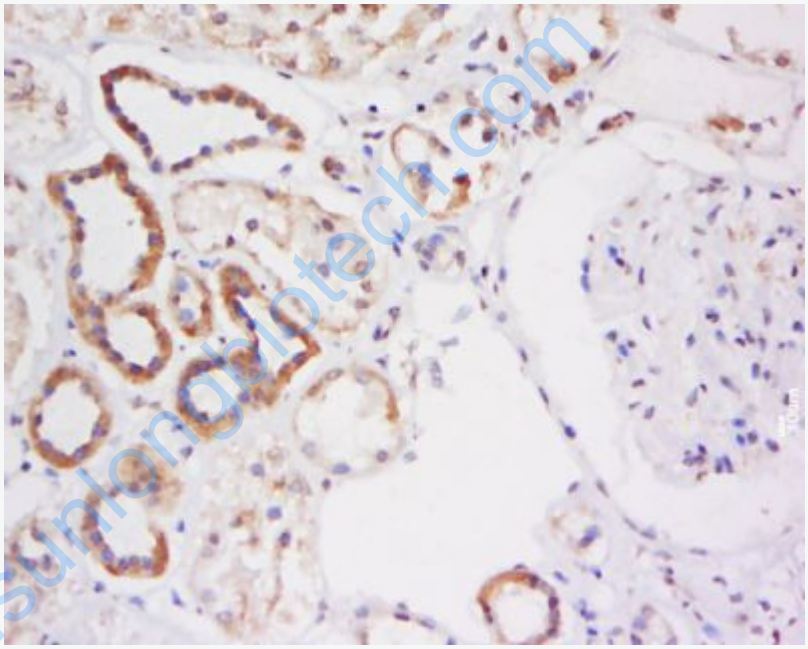
Entrez Gene: 6606 Human

Entrez Gene: 6607 Human

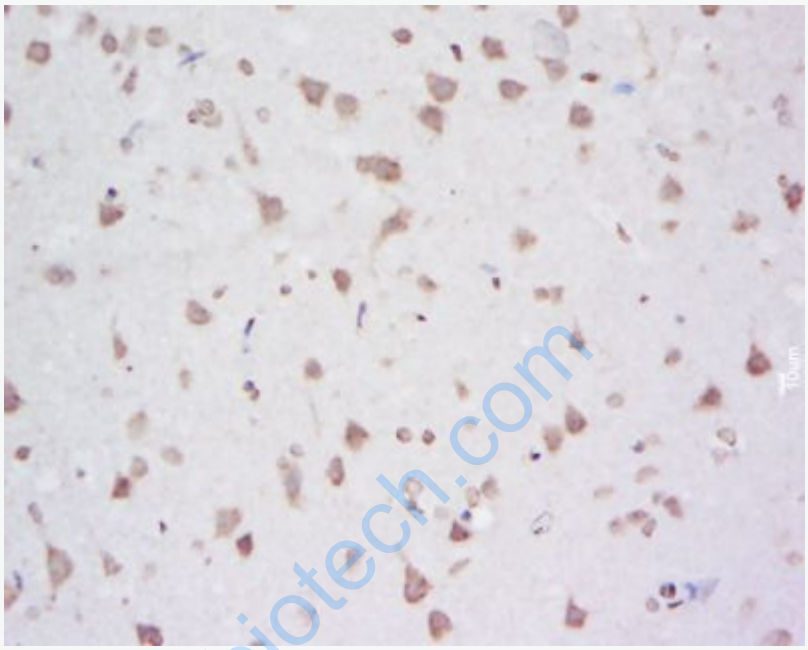
Omim: 600354 Human

Omim: 601627 Human

SwissProt: O14893 Human

	SwissProt: Q16637 Human Unigene: 202179 Human Unigene: 535788 Human **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Picture:	 Tissue/cell: Human kidney tissue; 4% Paraformaldehyde-fixed and paraffin-embedded; Antigen retrieval: citrate buffer (0.01M, pH 6.0), Boiling bathing for 15min; Block endogenous peroxidase by 3% Hydrogen peroxide for 30min; Blocking buffer (normal goat serum,C-0005) at 37℃ for 20 min; Incubation: Anti-SMN1 Polyclonal Antibody, Unconjugated(SL4628R) 1:200, overnight at 4°C, followed by conjugation to the secondary antibody(SP-0023) and

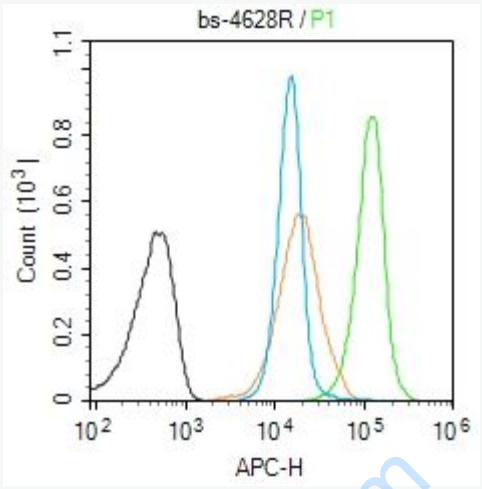
DAB(C-0010) staining



Tissue/cell: Rat brain tissue; 4% Paraformaldehyde-fixed and paraffin-embedded;

Antigen retrieval: citrate buffer (0.01M, pH 6.0), Boiling bathing for 15min; Block endogenous peroxidase by 3% Hydrogen peroxide for 30min; Blocking buffer (normal goat serum,C-0005) at 37℃ for 20 min;

Incubation: Anti-SMN1 Polyclonal Antibody, Unconjugated(SL4628R) 1:200, overnight at 4°C, followed by conjugation to the secondary antibody(SP-0023) and DAB(C-0010) staining



Blank control (Black line):Molt4 (Black).

Primary Antibody (green line): Rabbit Anti-SMN1 antibody (SL4628R)

Dilution: 1μg /10^6 cells;

Isotype Control Antibody (orange line): Rabbit IgG .

Secondary Antibody (white blue line): Goat anti-rabbit IgG-AF647

Dilution: 1μg /test.

Protocol

The cells were fixed with 4% PFA (10min at room temperature)and then permeabilized with 90% ice-cold methanol for 20 min at room temperature. The cells were then incubated in 5%BSA to block non-specific protein-protein interactions for 30 min at room temperature .Cells stained with Primary Antibody for 30 min at room temperature. The secondary antibody used for 40 min at room temperature. Acquisition of 20,000 events was performed.