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KO-Validated Anti-SMN1 Rabbit Monoclonal Antibody

Rabbit monoclonal antibody
Catalog # AGI2423

Product Information

Application	WB, FC, ICC
Primary Accession	Q16637
Reactivity	Human
Clonality	Monoclonal
Isotype	Rabbit IgG
Clone Names	25GB1840
Calculated MW	31849
Gene Name	SMN1
Aliases	SMN1; Survival Of Motor Neuron 1, Telomeric; SMNT; TDRD16A; Gemin-1; BCD541; GEMIN1; SMA1; SMA2; SMA3; Survival Motor Neuron Protein; Tudor Domain Containing 16A; Component Of Gems 1; SMA@; SMA; SMN; Spinal Muscular Atrophy (Werdnig-Hoffmann Disease, Kugelberg-Welander Disease); Survival Motor Neuron 1 Protein; T-BCD541; SMA4; SMNC
Immunogen	A synthesized peptide derived from human SMN1

Additional Information

Gene ID	6606;6607
Other Names	Survival motor neuron protein, Component of gems 1, Gemin-1, SMN1, SMN, SMNT

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:18984161, PubMed:9845364). 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:17178713, PubMed:21816274,

PubMed:22101937). 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).

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