
Product Name: KO-Validated SMN1 Recombinant Rabbit Monoclonal Antibody**Catalog #: KVA00116**

For research use only.

Summary

Description	KO&KD-Validated antibody
Host	Rabbit
Application	WB,FCM,ICC
Reactivity	Human,M
Conjugation	Unconjugated
Modification	Unmodified
Isotype	Rabbit IgG
Clonality	Rabbit mAb
Form	Liquid
Storage	Aliquot and store at -20°C (valid for 12 months). Avoid freeze/thaw cycles.
Shipping	Ice bags
Buffer	Supplied in PBS (pH 7.4) containing 50% glycerol, and 0.02% sodium azide.
Purification	Affinity purification

Application

Dilution Ratio	WB 1:1,000-1:5,000; FC 1:200-1:2,000; ICC 1:100-1:1,000
Molecular Weight	Calculated MW: 31.8kDa

Antigen Information

Gene Name	SMN1 SMN1; Survival Of Motor Neuron 1, Telomeric; SMNT; TDRD16A; Gemin-1; BCD541; GEMIN1; SMA1; SMA2; SMA3; Survival Motor Neuron Protein; Tudor Domain Containing 16A;
Alternative Names	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
Gene ID	6606.0
SwissProt ID	Q16637
Immunogen	A synthesized peptide derived from human SMN1

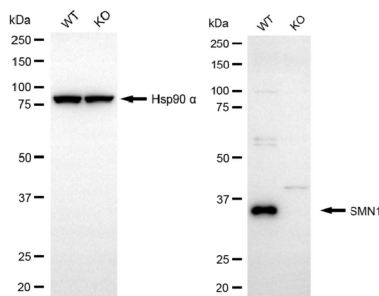
Background

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]

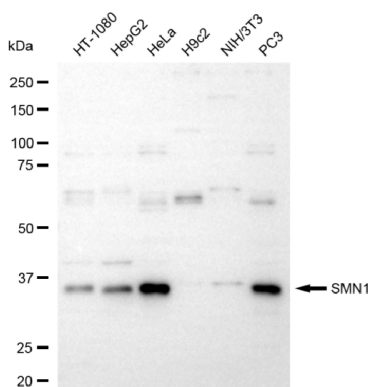
Research Area

Neuroscience, Epigenetics and Nuclear Signaling

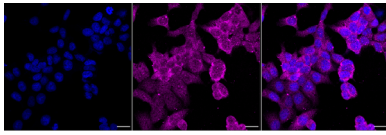
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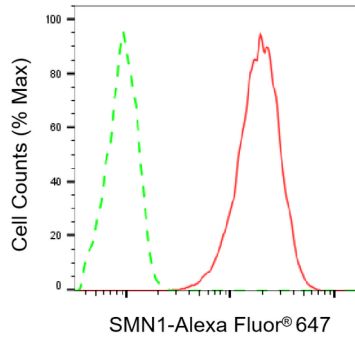
Western blotting analysis using SMN1 antibody (KVA00116). SMN1 expression in wild-type (WT) and SMN1 knockout (KO) HeLa cells with 20 µg of total cell lysates. Hsp90 α serves as a loading control. The blot was incubated with SMN1 antibody (KVA00116, 1:5,000) and HRP-conjugated goat anti-rabbit secondary antibody (APS0635, 1:10,000) respectively.



Western blotting analysis using SMN1 antibody (KVA00116). Total cell lysates (30 µg) from various cell lines were loaded and separated by SDS-PAGE. The blot was incubated with SMN1 antibody (KVA00116, 1:5,000) and HRP-conjugated goat anti-rabbit secondary antibody (APS0635, 1:10,000) respectively.



Immunocytochemical staining of HAP-1 cells with SMN1 antibody (KVA00116, 1:1,000). Nuclei were stained blue with DAPI; SMN1 was stained magenta with Alexa Fluor® 647. Images were taken using Leica stellaris 5. Protein abundance based on laser Intensity and smart gain: Medium. Scale bar, 20 μ m.



Flow cytometric analysis of SMN1 expression in HAP-1 cells using SMN1 antibody (KVA00116, 1:2,000). Green, isotype control; red, SMN1.