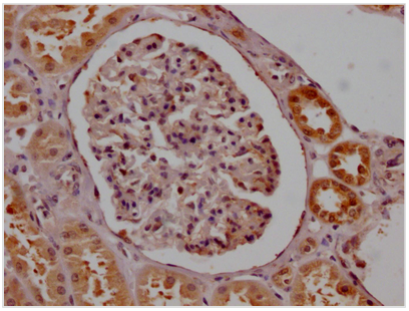
SMN1 Antibody

Product Code	CSB-RA567382A0HU
Storage	Upon receipt, store at -20°C or -80°C. Avoid repeated freeze.
Uniprot No.	Q16637
Immunogen	A synthesized peptide derived from human SMN1
Species Reactivity	Human
Tested Applications	ELISA, IHC; Recommended dilution: IHC:1:50-1:200
Relevance	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 pre-mRNAs. 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 pICln-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).
Form	Liquid
Conjugate	Non-conjugated
Storage Buffer	Rabbit IgG in phosphate buffered saline, pH 7.4, 150mM NaCl, 0.02% sodium azide and 50% glycerol.
Purification Method	Affinity-chromatography
Isotype	Rabbit IgG
Clonality	Monoclonal
Product Type	Recombinant Antibody
Immunogen Species	Homo sapiens (Human)
Research Area	Epigenetics and Nuclear Signaling; Neuroscience; Signal transduction
Gene Names	SMN1
Accession NO.	8B10
Image	



IHC image of CSB-RA567382A0HU diluted at 1:100 and staining in paraffin-embedded human kidney tissue performed on a Leica BondTM system. After dewaxing and hydration, antigen retrieval was mediated by high pressure in a citrate buffer (pH 6.0). Section was blocked with 10% normal goat serum 30min at RT. Then primary antibody (1% BSA) was incubated at 4°C overnight. The primary is detected by a Goat anti-rabbit IgG polymer labeled by HRP and visualized using 0.05% DAB.

Description

The SMN1 encodes the survival motor neuron (SMN) protein, which in conjunction with several Gemin proteins forms an SMN complex whose chaperone function facilitates the assembly of spliceosomal snRNP particles, essential components of the spliceosome complex, and hence plays a critical role in pre-mRNA splicing. Loss or mutation of the SMN1 gene causes spinal muscular atrophy (SMA), an autosomal recessive neuromuscular disease with α-motor neuron dysfunction and muscular atrophy. SMN is also implicated in mRNA transport in the axon of the nerve, the disturbance of which might explain the vulnerability of motor neurons in SMA.

This recombinant SMN1 antibody was developed with the Single B cell platform. The main process included identification and isolation of single B cells; amplification and cloning of SMN1 antibody gene; expression, screening, and identification of antibody specificity. And this SMN1 antibody has been validated in ELISA, IHC.