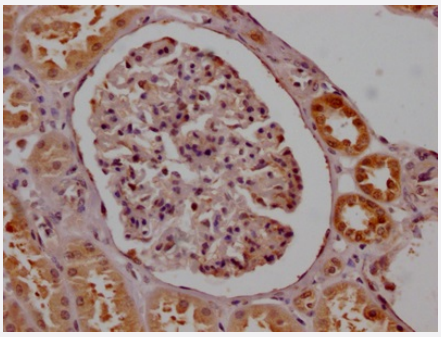
RecomAb™

SMN1 recombinant monoclonal antibody, clone 8B10

Catalog # RAB07471 Size 100 uL

Applications



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections)

Immunohistochemical analysis of paraffin-embedded human kidney tissue using SMN1 recombinant monoclonal antibody, clone 8B10 (Cat # RAB07471) 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.

Specification

Product Description	Rabbit recombinant monoclonal antibody raised against human SMN1.
Antibody Species	Rabbit
Immunogen	Original antibody is raised against a synthetic peptide corresponding to human SMN1.
Reactivity	Human
Form	Liquid
Purification	Affinity chromatography purification
Isotype	IgG
Recommend Usage	ELISA Immunohistochemistry (1:50-1:200) The optimal working dilution should be determined by the end user.
Storage Buffer	In PBS, pH7.4 (150 mM NaCl, 0.02% sodium azide and 50% glycerol)

Storage Instruction	Store at -20°C or -80°C. Aliquot to avoid repeated freezing and thawing.
Note	This product contains sodium azide: a POISONOUS AND HAZARDOUS SUBSTANCE which shoul d be handled by trained staff only.

Applications

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- Enzyme-linked Immunoabsorbent Assay

Gene Info — SMN1

Entrez GeneID	6606
Protein Accession#	Q16637
Gene Name	SMN1
Gene Alias	BCD541, SMA, SMA1, SMA2, SMA3, SMA4, SMA@, SMN, SMNT, T-BCD541
Gene Description	survival of motor neuron 1, telomeric
Omim ID	253300 253400 253550 271150 600354
Gene Ontology	Hyperlink

Gene Summary	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. Two transcript variants encoding distinct isoforms have been described. [provided by RefSeq
Other Designations	OTTHUMP00000125198|gemin 1

Disease

- Acute Disease
- Amyotrophic lateral sclerosis
- Chronic Disease
- Disease Progression
- Genetic Predisposition to Disease
- Muscular Atrophy
- Nerve Degeneration
- Spinal Muscular Atrophies of Childhood
- Spinal muscular atrophy