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[Primary Antibody]

Rabbit Anti-Gemin 1 antibody

Catalog Number: SL11561R

FOR RESEARCH USE ONLY

[DATASHEET]

Host:	Rabbit
Isotype:	IgG
Clonality:	Polyclonal
Target:	Gemin 1
Immunogen:	KLH conjugated synthetic peptide derived from human Gemin 1: 31-100/294
Purification:	affinity purified by Protein A
Concentration:	1mg/ml
Storage:	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol. Shipped at 4℃. Store at -20 °C for one year. Avoid repeated freeze/thaw cycles.
Applications:	WB=1:500-2000 IHC-P=1:100-500 IHC-F=1:100-500 IF=1:100-500 （Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Reactivity:	Mouse, (predicted: Human, Rat, Dog, Pig, Cow, Rabbit)
Predicted MW.:	31kDa
Subcellular Location:	The nucleus cytoplasmic
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

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	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, Sep 2008] **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: Defects in SMN1 are the cause of spinal muscular atrophy autosomal recessive type 1 (SMA1) [MIM:253300]. Spinal muscular atrophy refers to a group of neuromuscular disorders 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

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	age. SMA1 patients never achieve the ability to sit. Defects in SMN1 are the cause of spinal muscular atrophy autosomal recessive type 2 (SMA2) [MIM:253550]. SMA2 is an autosomal recessive spinal muscular atrophy of intermediate severity, with onset between 6 and 18 months. Patients do not reach the motor milestone of standing, and survive into adulthood. Defects in SMN1 are the cause of spinal muscular atrophy autosomal recessive type 3 (SMA3) [MIM:253400]. SMA3 is an autosomal recessive spinal muscular atrophy with onset after 18 months. SMA3 patients develop ability to stand and walk and survive into adulthood. Defects in SMN1 are the cause of spinal muscular atrophy autosomal recessive type 4 (SMA4) [MIM:271150]. SMA4 is an autosomal recessive spinal muscular atrophy characterized by symmetric proximal muscle weakness with onset in adulthood and slow disease progression. SMA4 patients can stand and walk. **Similarity:** Belongs to the SMN family. Contains 1 Tudor domain. **Database links:** **Entrez Gene:** 6606 Human **Entrez Gene:** 6607 Human **Entrez Gene:** 20595 Mouse **Entrez Gene:** 64301 Rat Omim: 600354 Human **SwissProt:** Q16637 Human **SwissProt:** P97801 Mouse **SwissProt:** O35876 Rat **Unigene:** 202179 Human **Unigene:** 535788 Human **Unigene:** 2025 Mouse **Unigene:** 1119 Rat

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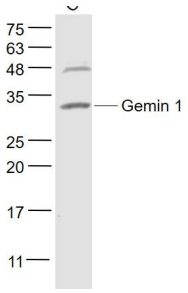
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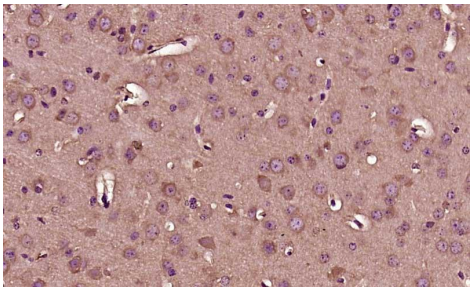
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[VALIDATION IMAGES]





Sample:
Cerebrum (Mouse) Lysate at 40 ug
Primary: Anti- Gemin 1 (SL11561R) at 1/1000 dilution
Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution
Predicted band size: 31 kD
Observed band size: 31 kD

Paraformaldehyde-fixed, paraffin embedded (Mouse brain); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (Gemin 1) Polyclonal Antibody, Unconjugated (SL11561R) at 1:400 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.

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