SZABO SCANDIC

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# Produktinformation

- Forschungsprodukte & Biochemikalien
- Zellkultur & Verbrauchsmaterial
- Diagnostik & molekulare Diagnostik
- Laborgeräte & Service

Weitere Information auf den folgenden Seiten!
See the following pages for more information!

## Lieferung & Zahlungsart

siehe unsere Liefer- und Versandbedingungen

## Zuschläge

- Mindermengenzuschlag
- Trockeneiszuschlag
- Gefahrgutzuschlag
- Expressversand


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# Datasheet

## SMN1 recombinant monoclonal antibody, clone R05-3B5

**Catalog Number:** RAB02258

**Regulatory Status:** For research use only (RUO)

**Product Description:** Rabbit recombinant monoclonal antibody raised against human SMN1.

**Clone Name:** R05-3B5

**Immunogen:** Original antibody is raised against a synthetic peptide corresponding to human SMN1.

**Theoretical MW (kDa):** Calculated MW: 32 kD

**Antibody Species:** Rabbit

**Protocols:** See our web site at http://www.abnova.com/support/protocols.asp or product page for detailed protocols

**Form:** Liquid

**Purification:** Affinity purification

**Isotype:** IgG

**Recommend Usage:** Immunofluorescence (1:50-1:200)
Western Blot (1:500-1:1000)
The optimal working dilution should be determined by the end user.

**Storage Buffer:** In 50 mM Tris-Glycine, pH 7.4 (0.15 M NaCl, 40% Glycerol, 0.01% Sodium azide and 0.05% BSA)

**Storage Instruction:** Store at -20 °C.
Aliquot to avoid repeated freezing and thawing.

**Entrez GeneID:** 6606

**Gene Symbol:** SMN1

**Gene Alias:** BCD541, SMA, SMA1, SMA2, SMA3, SMA4, SMA@, SMN, SMNT, T-BCD541

**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]