

## Product Information

### Anti-SMN antibody, Mouse monoclonal

clone 2B1, purified from hybridoma cell culture

Catalog Number **S2944**

Synonym: Anti-Survival of Motor Neurons

#### Product Description

Monoclonal Anti-SMN, Clone 2B1 (mouse IgG1 isotype) is derived from the 2B1 hybridoma produced by the fusion of mouse myeloma cells and splenocytes from mice immunized with recombinant human SMN.<sup>1</sup> The isotype is determined using a double diffusion immunoassay and Mouse Monoclonal Antibody Isotyping Reagents, Catalog Number ISO2.

The antibody recognizes human,<sup>1</sup> mouse, and *Xenopus* SMN. The antibody may be used in immunoblotting (~35 kDa),<sup>1, 2</sup> immunoprecipitation,<sup>2</sup> and immunocytochemistry.<sup>2</sup>

Spinal muscular atrophy (SMA) is caused by reduced expression or mutations in the Survival of Motor Neurons (SMN) protein. Two SMN proteins (SMN1 and SMN2) are encoded in human chromosome 5. Deletion or mutation in the telomeric copy (SMN1) causes the SMA phenotype. The severity of SMA is in direct correlation with the expression level of the SMN protein, either from the SMN1 gene or a different spliced form of SMN from the SMN2 gene.<sup>3-4</sup>

The SMN protein forms a multi-protein complex with the Gemin proteins. Gemin2, 3, 5 and 7 interact directly with the SMN protein while the interaction of Gemin4 and 6 requires Gemin3 and 7, respectively. The SMN complex interacts with various protein substrates such as Sm and Lsm proteins of the spliceosomal snRNPs, fibrillarin, GAR1, RNA helicase A, the human hnRNP proteins (hnRNPQ, U and R), coilin and p53. Arginine- and glycine-rich regions in the protein substrates are important for the interaction with the SMN complex. Furthermore, methylation of arginines in these regions by the methylome (aka PRMT5) enhances their affinity for SMN. The SMN complex is important in various biological processes such as assembly and restructuring of spliceosomal snRNPs, pre mRNA splicing and transcription.<sup>5,6</sup>

Monoclonal antibodies specific for SMN are an important tool for studying the role of the SMN protein in nuclear processes.

#### Reagent

Supplied as a solution in 0.01M PBS, pH 7.4, containing 15 mM sodium azide as a preservative. Antibody concentration: ~2 mg/ml.

#### Precautions and Disclaimer

This product is for R&D use only, not for drug, household, or other uses.

#### Storage/Stability

For continuous use, store at 2-8 °C for up to one month. For extended storage, freeze in working aliquots. Repeated freezing and thawing, or storage in "frost-free" freezers, is not recommended. If slight turbidity occurs upon prolonged storage, clarify the solution by centrifugation before use. Working dilution samples should be discarded if not used within 12 hours.

#### Product Profile

Immunoblotting: a working concentration of 2-4 µg/mL is determined using A431 cell extract.

**Note:** In order to obtain best results in different techniques and preparations we recommend determining optimal working dilution by titration test.

#### References

1. Liu, Q., et al., *EMBO J.*, **15**, 3555-3565 (1996).
2. Charroux, B., et al., *J. Cell Biol.*, **147**, 1181-1193 (1999).
3. Paushkin, S., et al., *Curr. Opin. Cell Biol.*, **14**, 305-312 (2002).
4. Pellizzoni, L., et al., *Proc. Natl. Acad. Sci. USA*, **96**, 11167-11172 (1999).
5. Baccon, J., et al., *J. Biol. Chem.*, **277**, 31957-31962 (2002).
6. Pellizzoni, L., *Science*, **298**, 1775-1779 (2002).

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