SMN1 monoclonal antibody (2B1)

Survival Motor Neuron (SMN) is a ~38 kDa protein produced chiefly by the SMN1 gene, located on the telomeric portion of chromosome 5q. A nearly identical centromeric copy of the gene (SMN2) also produces a small amount of full-length SMN protein, but due to a translationally silent C(R)T transition that results in alternative splicing of the pre-mRNA, most of the resulting SMN is truncated, causing reduced protein stability and lower overall SMN levels. Deletion or mutation of the SMN1 gene results in a reduced level of full-length SMN protein and manifests as a range of neuromuscular phenotypes in humans as the disease spinal muscular atrophy (SMA). SMA is characterized by muscle weakness and atrophy, functional disability and is the most common lethal genetic disease of infants and toddlers. Approximately one in 35 adults is a carrier of the SMN1 mutation. The incidence of SMA is 1 in 6,000 to 1 in 10,000 live births. SMN protein is present in the cell cytoplasm, and also in the nucleus where it is concentrated in “gem” structures associated with Cajal bodies. SMN protein is a constituent of Gemin-containing complexes, and is thought to participate in many aspects of RNA metabolism. SMN complexes have been shown to mediate the assembly of uridine-rich small nuclear ribonucleoproteins (snRNPs), which in turn act as critical components of spliceosomes.

This antibody is covered by our Worry-Free Guarantee.

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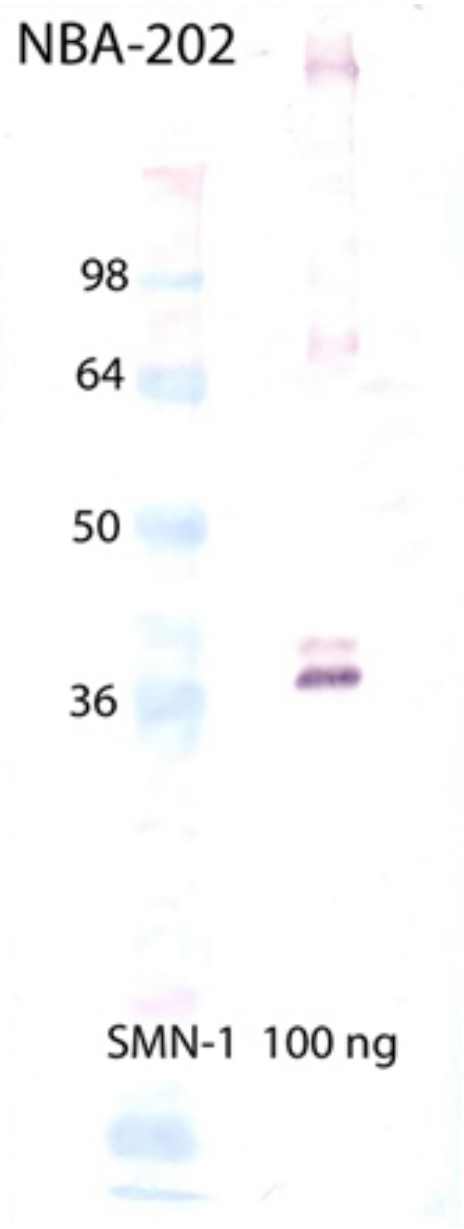
ADI-NBA-202-200	200µg

Manuals, SDS & CofA View Online »



Western Blot Analysis of SMN1, mAb (2B1) (Prod. No. NBA-202). Lane 1: MWM, Lane 2: Empty; and Lane 3: SMN1 (human), (recombinant) (His-tag) (Prod. No. NBP-201).

Handling & Storage

Handling	Avoid freeze/thaw cycles.
Long Term Storage	-20°C
Shipping	Blue Ice

Regulatory Status

RUO - Research Use Only

Product Details

Alternative Name	Survival Motor Neuron
Application	WB
Application Notes	Detects a band of ~38kDa by Western blot.
Clone	2B1
Formulation	Liquid. In PBS, pH 7.2, containing 0.09% sodium azide.
Host	Mouse
Immunogen	Recombinant fragment corresponding to the carboxy-terminus of human SMN1.
Isotype	IgG
Purity Detail	Protein G-affinity purified.
Recommendation Dilutions/Conditions	Western Blot (1:1,000, colorimetric)Suggested dilutions/conditions may not be available for all applications.Optimal conditions must be determined individually for each application.
Source	Purified from hybridoma tissue culture supernatant.
Species Reactivity	Human
UniProt ID	Q16637

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