

Product datasheet

Anti-SMN/Gemin 1 antibody [EPR4430] ab108424

Recombinant RabMAb

2 Images

Overview

Product name	Anti-SMN/Gemin 1 antibody [EPR4430]
Description	Rabbit monoclonal [EPR4430] to SMN/Gemin 1
Host species	Rabbit
Tested applications	Suitable for: WB, IP, IHC-P Unsuitable for: Flow Cyt or ICC
Species reactivity	Reacts with: Mouse, Rat, Human
Immunogen	Synthetic peptide within Human SMN/Gemin 1 aa 150-250. The exact sequence is proprietary.
Positive control	HeLa, HepG2, K562 cell lysates
General notes	

Our RabMAb[®] technology is a patented hybridoma-based technology for making rabbit monoclonal antibodies. For details on our patents, please refer to [RabMab[®] patents](#)

This product is a recombinant rabbit monoclonal antibody.

Properties

Form	Liquid
Storage instructions	Shipped at 4°C. Store at -20°C. Stable for 12 months at -20°C.
Storage buffer	pH: 7.20 Preservative: 0.05% Sodium azide Constituents: 0.1% BSA, 40% Glycerol, 9.85% Tris glycine, 50% Tissue culture supernatant
Purity	Tissue culture supernatant
Clonality	Monoclonal
Clone number	EPR4430
Isotype	IgG

Applications

Our [Abpromise guarantee](#) covers the use of **ab108424** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Application	Abreviews	Notes
WB		1/1000 - 1/10000. Predicted molecular weight: 32 kDa.
IP		1/10 - 1/100.
IHC-P		1/500.

Application notes Is unsuitable for Flow Cyt or ICC.

Target

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.

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).

Involvement in 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 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.

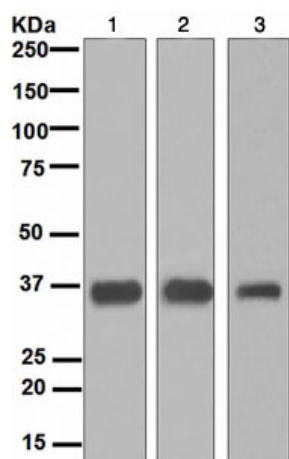
Defects in SMN1 are the cause of spinal muscular atrophy autosomal recessive type 3 (SMA3) [MIM:253400]. SMA3 is an autosomal recessive spinal muscular atrophy with onset after 18 months. SMA3 patients develop ability to stand and walk and survive into adulthood.

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.

Sequence similarities Belongs to the SMN family.
Contains 1 Tudor domain.

Cellular localization Cytoplasm. Nucleus > gem. Localized in subnuclear structures next to coiled bodies, called Gemini or Cajal bodies.

Images



Western blot - Anti-SMN/Gemin 1 antibody
[EPR4430] (ab108424)

All lanes : Anti-SMN/Gemin 1 antibody
[EPR4430] (ab108424) at 1/1000 dilution

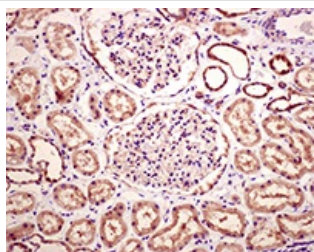
Lane 1 : HeLa cell lysate

Lane 2 : HepG2 cell lysate

Lane 3 : K562 cell lysate

Lysates/proteins at 10 µg per lane.

Predicted band size: 32 kDa



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-SMN/Gemin 1 antibody
[EPR4430] (ab108424)

ab108424 staining Gemin 1 in paraffin-embedded Human kidney tissue by Immunohistochemistry at dilution of 1:500.

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