abcam

Product datasheet

Anti-SMN/Gemin 1 antibody ab223068

3 References 3 Images

Overview

Product name Anti-SMN/Gemin 1 antibody

Description Rabbit polyclonal to SMN/Gemin 1

Host species Rabbit

Tested applications Suitable for: WB, IHC-P, ICC/IF

Species reactivity Reacts with: Human

Predicted to work with: Macaque monkey

Immunogen Recombinant fragment corresponding to Human SMN/Gemin 1 aa 1-200.

Database link: Q16637

Run BLAST with
Run BLAST with

Positive control WB: A549 and HeLa whole cell lysate. IHC: Human testis tissue. ICC/IF: HepG2 cells.

General notesThe Life Science industry has been in the grips of a reproducibility crisis for a number of years.

Abcam is leading the way in addressing this with our range of recombinant monoclonal antibodies and knockout edited cell lines for gold-standard validation. Please check that this product meets

your needs before purchasing.

If you have any questions, special requirements or concerns, please send us an inquiry and/or contact our Support team ahead of purchase. Recommended alternatives for this product can be

found below, along with publications, customer reviews and Q&As

Properties

Form Liquid

Storage instructions Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C long

term. Avoid freeze / thaw cycle.

Storage buffer pH: 7.40

Preservative: 0.03% Proclin 300

Constituents: 50% Glycerol (glycerin, glycerine), PBS

Purity Protein G purified

Purification notes Purity >95%

Clonality Polyclonal

Isotype IgG

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Applications

The Abpromise guarantee

Our Abpromise quarantee covers the use of ab223068 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		1/500 - 1/5000.
IHC-P		1/20 - 1/200.
ICC/IF		1/50 - 1/200.

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

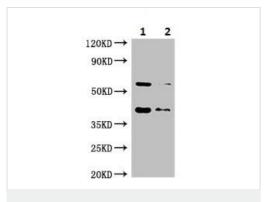
symmetric proximal muscle weakness with onset in adulthood and slow disease progression. SMA4 patients can stand and walk.

Sequence similarities

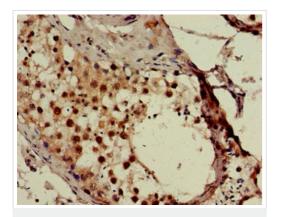
Belongs to the SMN family. Contains 1 Tudor domain.

Cellular localization

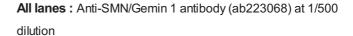
Cytoplasm. Nucleus > gem. Localized in subnuclear structures next to coiled bodies, called Gemini of Cajal bodies.



Western blot - Anti-SMN/Gemin 1 antibody (ab223068)



Immunohistochemistry (Formalin/PFA-fixed paraffinembedded sections) - Anti-SMN/Gemin 1 antibody (ab223068)



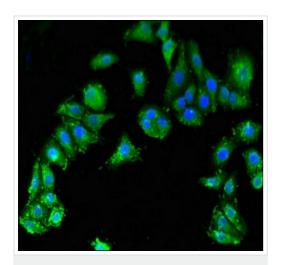
Lane 1 : A549 (human lung carcinoma cell line) whole cell lysateLane 2 : HeLa (human epithelial cell line from cervix adenocarcinoma) whole cell lysate

Secondary

All lanes: Goat polyclonal to rabbit IgG at 1/50000 dilution

Developed using the ECL technique.

Paraffin-embedded human testis tissue stained for SMN/Gemin 1 using ab223068 at 1/100 dilution in immunohistochemical analysis.



Immunocytochemistry/ Immunofluorescence - Anti-SMN/Gemin 1 antibody (ab223068)

PFA-fixed, Triton X-100 permeabilized HepG2 (human liver hepatocellular carcinoma cell line) cells stained for SMN/Gemin 1(green) using ab223068 at 1/100 dilution in ICC/IF. Secondary: Alexa Fluor 488® conjugated Goat Anti-Rabbit IgG (H+L).

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