abcam

Product datasheet

Anti-SMN/Gemin 1 antibody ab88979

1 References 2 图像

概述

产品名称	Anti-SMN/Gemin 1**抗体**
描述	**小鼠多克隆抗体**to SMN/Gemin 1
宿主	Mouse
经测试应用	**适用于:** WB
种属反应性	**与反应:** Human
免疫原	Recombinant full length protein within Human SMN/Gemin 1 aa 1-300. The exact immunogen sequence used to generate this antibody is proprietary information. If additional detail on the immunogen is needed to determine the suitability of the antibody for your needs, please **contact** our Scientific Support team to discuss your requirements.
阳性对照	Human kidney lysate
常规说明	The 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

性能

形式	Liquid
存放说明	Shipped at 4°C. Upon delivery aliquot and store at -20°C or -80°C. Avoid repeated freeze / thaw cycles.
存储溶液	pH: 7.40 Constituent: PBS
纯度	Protein A purified
克隆	**多克隆**
同种型	IgG

应用

The Abpromise guarantee **Abpromise™承诺保证使用ab88979于以下的经测试应用**

"应用说明"部分 下显示的仅为推荐的起始稀释度;实际最佳的稀释度/浓度应由使用者检定。

应用	Ab评论	说明
WB		1/500 - 1/1000. Predicted molecular weight: 31 kDa.

靶标

功能

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.

组织特异性

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).

疾病相关

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.

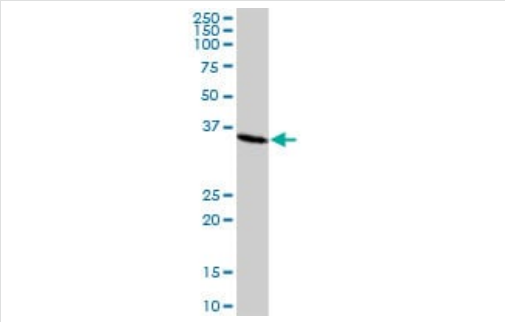
序列相似性

Belongs to the SMN family.
Contains 1 Tudor domain.

细胞定位

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

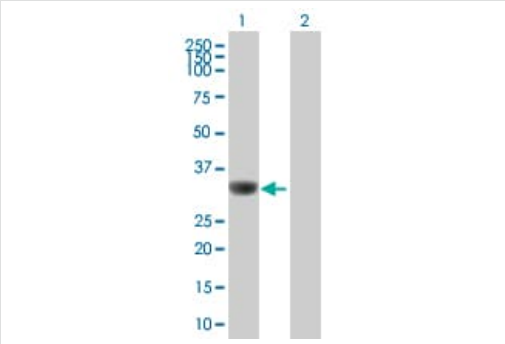
图片



Western blot - Anti-SMN/Gemin 1 antibody (ab88979)

Anti-SMN/Gemin 1 antibody (ab88979) at 1/500 dilution + Human kidney lysate at 50 µg

Predicted band size: 31 kDa



Western blot - Anti-SMN/Gemin 1 antibody (ab88979)

All lanes : Anti-SMN/Gemin 1 antibody (ab88979) at 1/500 dilution

Lane 1 : SMN2-transfected 293T cell lysate
Lane 2 : untransfected 293T cell lysate

Lysates/proteins at 25 µg per lane.

Predicted band size: 31 kDa
Observed band size: 31 kDa

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