神经鞘脂激活蛋白 3 抗体

产品货号： mlR13452

英文名称： GM2A

中文名称： 神经鞘脂激活蛋白 3 抗体

别　　名： Cerebroside sulfate activator protein; Ganglioside GM2 activator isoform short; Ganglioside GM2 activator precursor; GM2 AP; GM2 ganglioside activator; GM2 ganglioside activator protein; GM2-AP; GM2A; GM2AP; OTTHUMP00000160619; SAP 3; SAP-3; SAP3; SAP3_HUMAN; Shingolipid activator protein 3; Sphingolipid activator protein 3.

研究领域： 肿瘤　细胞生物　神经生物学　细胞类型标志物　新陈代谢

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应： Human, Rat,

产品应用： WB=1:500-2000 ELISA=1:500-1000 IHC-P=1:400-800 IHC-F=1:400-800 ICC=1:100-500 IF=1:100-500（石蜡切片需做抗原修复）
not yet tested in other applications.
optimal dilutions/concentrations should be determined by the end user.

分 子 量： 17/18kDa

细胞定位： 细胞浆

性　　状： Lyophilized or Liquid

浓　　度： 1mg/ml

免 疫 原： KLH conjugated synthetic peptide derived from human GM2A/SAP3:131-193/193

亚　　型： IgG

纯化方法： affinity purified by Protein A

储 存 液： 0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.

保存条件： Store at -20 ˚C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20˚C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 ˚C.

PubMed： PubMed

产品介绍： This gene encodes a small glycolipid transport protein which acts as a substrate specific co-factor for the lysosomal enzyme beta-hexosaminidase A. Beta-hexosaminidase A, together with GM2 ganglioside activator, catalyzes the degradation of the ganglioside GM2, and other molecules containing terminal N-acetyl hexosamines. Mutations in this gene result in GM2-gangliosidosis type AB or the AB variant of Tay-Sachs disease. Alternative splicing results in multiple transcript variants. [provided by RefSeq, Nov 2009].

Function:

Binds gangliosides and stimulates ganglioside GM2 degradation. It stimulates only the breakdown of ganglioside GM2 and glycolipid GA2 by beta-hexosaminidase A. It extracts single GM2 molecules from membranes and presents them in soluble form to beta-hexosaminidase A for cleavage of N-acetyl-D-galactosamine and conversion to GM3.

Subcellular Location:

Lysosome.

Post-translational modifications:

The serines in positions 32 and 33 are absent in 80% of the sequenced protein.

DISEASE:

Defects in GM2A are the cause of GM2-gangliosidosis type AB (GM2GAB) [MIM:272750]; also known as Tay-Sachs disease AB variant. GM2-gangliosidosis is an autosomal recessive lysosomal storage disease marked by the accumulation of GM2 gangliosides in the neuronal cells. GM2GAB is characterized by GM2 gangliosides accumulation in the presence of both hexosaminidase A and B.

SWISS:

P17900

Gene ID:

2760

Important Note:

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

产品图片