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Rabbit Anti-GM2A antibody

SL13452R

Product Name:	GM2A
Chinese Name:	神经鞘脂激活蛋白3抗体
Alias:	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.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Rat,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	17/18kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human GM2A/SAP3:131-193/193
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	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
Product Detail:	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

Database links:

Entrez Gene: 2760Human

Entrez Gene: 14667Mouse

Entrez Gene: 282838Rat

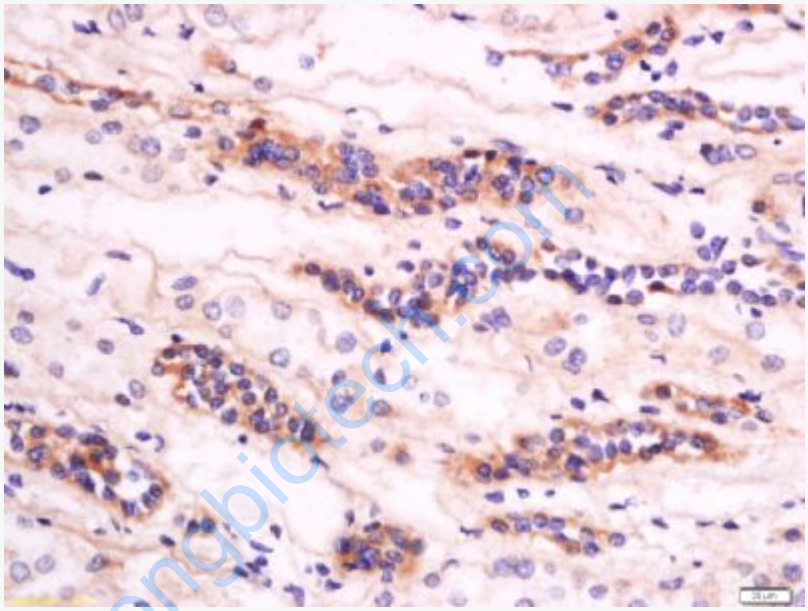
Omim: 613109Human

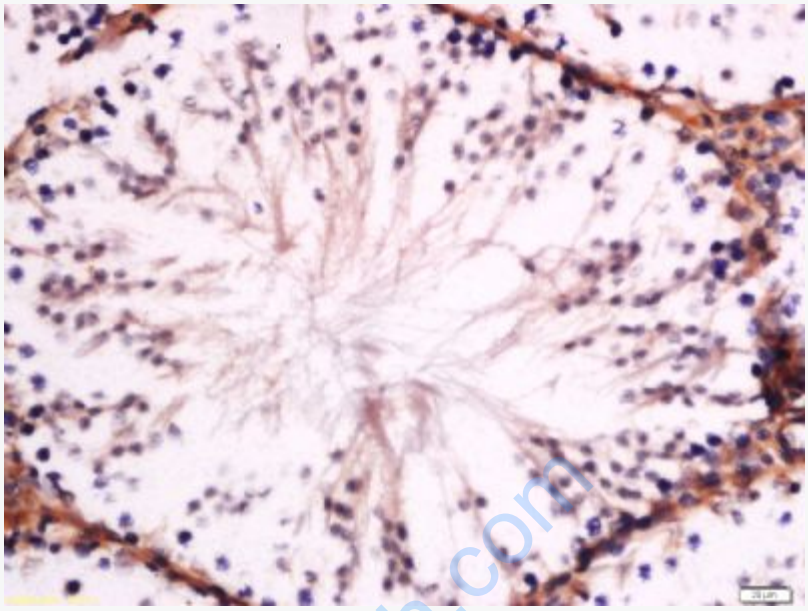
SwissProt: P17900Human

SwissProt: Q60648Mouse

Unigene: 483873Human

Unigene: 287807Mouse

	Unigene: 98783Rat **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Picture:	 Tissue/cell: human kidney tissue; 4% Paraformaldehyde-fixed and paraffin-embedded; Antigen retrieval: citrate buffer (0.01M, pH 6.0), Boiling bathing for 15min; Block endogenous peroxidase by 3% Hydrogen peroxide for 30min; Blocking buffer (normal goat serum,C-0005) at 37℃ for 20 min; Incubation: Anti-GM2A Polyclonal Antibody, Unconjugated(SL13452R) 1:200, overnight at 4°C, followed by conjugation to the secondary antibody(SP-0023) and DAB(C-0010) staining



Tissue/cell: Rat testis tissue; 4% Paraformaldehyde-fixed and paraffin-embedded;

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