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Rabbit Anti-Acid sphingomyelinase antibody

SL6318R

Product Name:	Acid sphingomyelinase
Chinese Name:	酸性神经鞘磷脂酶抗体
Alias:	Acid sphingomyelinase; ASM; ASM_HUMAN; aSMase; NPD; Smpd1; Sphingomyelin phosphodiesterase 1 acid lysosomal; Sphingomyelin phosphodiesterase.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Pig,Cow,Rabbit,
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:	64kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human Acid sphingomyelinase:201-300/629
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:	Converts sphingomyelin to ceramide. Also has phospholipase C activities toward 1,2-diacylglycerolphosphocholine and 1,2-diacylglycerolphosphoglycerol. Isoform 2 and isoform 3 have lost catalytic activity. Involvement in disease: Defects in SMPD1 are the cause of Niemann-Pick disease type A (NPDA) ; also known as Niemann-Pick disease classical infantile form. It is an early-

	onset lysosomal storage disorder caused by failure to hydrolyze sphingomyelin to ceramide. It results in the accumulation of sphingomyelin and other metabolically related lipids in reticuloendothelial and other cell types throughout the body, leading to cell death. Niemann-Pick disease type A is a primarily neurodegenerative disorder characterized by onset within the first year of life, mental retardation, digestive disorders, failure to thrive, major hepatosplenomegaly, and severe neurologic symptoms. The severe neurological disorders and pulmonary infections lead to an early death, often around the age of four. Clinical features are variable. A phenotypic continuum exists between type A (basic neurovisceral) and type B (purely visceral) forms of Niemann-Pick disease, and the intermediate types encompass a cluster of variants combining clinical features of both types A and B. **Function:** Converts sphingomyelin to ceramide. Also has phospholipase C activities toward 1,2-diacylglycerolphosphocholine and 1,2-diacylglycerolphosphoglycerol. Isoform 2 and isoform 3 have lost catalytic activity. **Subunit:** Monomer. **Subcellular Location:** Lysosome. **DISEASE:** Defects in SMPD1 are the cause of Niemann-Pick disease type A (NPDA) [MIM:257200]; also known as Niemann-Pick disease classical infantile form. It is an early-onset lysosomal storage disorder caused by failure to hydrolyze sphingomyelin to ceramide. It results in the accumulation of sphingomyelin and other metabolically related lipids in reticuloendothelial and other cell types throughout the body, leading to cell death. Niemann-Pick disease type A is a primarily neurodegenerative disorder characterized by onset within the first year of life, mental retardation, digestive disorders, failure to thrive, major hepatosplenomegaly, and severe neurologic symptoms. The severe neurological disorders and pulmonary infections lead to an early death, often around the age of four. Clinical features are variable. A phenotypic continuum exists between type A (basic neurovisceral) and type B (purely visceral) forms of Niemann-Pick disease, and the intermediate types encompass a cluster of variants combining clinical features of both types A and B. Defects in SMPD1 are the cause of Niemann-Pick disease type B (NPDB) [MIM:607616]; also known as Niemann-Pick disease visceral form. It is a late-onset lysosomal storage disorder caused by failure to hydrolyze sphingomyelin to ceramide. It results in the accumulation of sphingomyelin and other metabolically related lipids in reticuloendothelial and other cell types throughout the body, leading to cell death. Clinical signs involve only visceral organs. The most constant sign is hepatosplenomegaly which can be associated with pulmonary symptoms. Patients remain free of neurologic manifestations. However, a phenotypic continuum exists between type A (basic neurovisceral) and type B (purely visceral) forms of Niemann-

Pick disease, and the intermediate types encompass a cluster of variants combining clinical features of both types A and B. In Niemann-Pick disease type B, onset of the first symptoms occurs in early childhood and patients can survive into adulthood.

Similarity:
Belongs to the acid sphingomyelinase family.
Contains 1 saposin B-type domain.

SWISS:
P17405

Gene ID:
6609

Database links:

Entrez Gene: 505097Cow

Entrez Gene: 485334Dog

Entrez Gene: 100720041Guinea pig

Entrez Gene: 6609Human

Entrez Gene: 20597Mouse

Entrez Gene: 100353898Rabbit

Entrez Gene: 308909Rat

Omim: 607608Human

SwissProt: Q0VD19Cow

SwissProt: P17405Human

SwissProt: Q04519Mouse

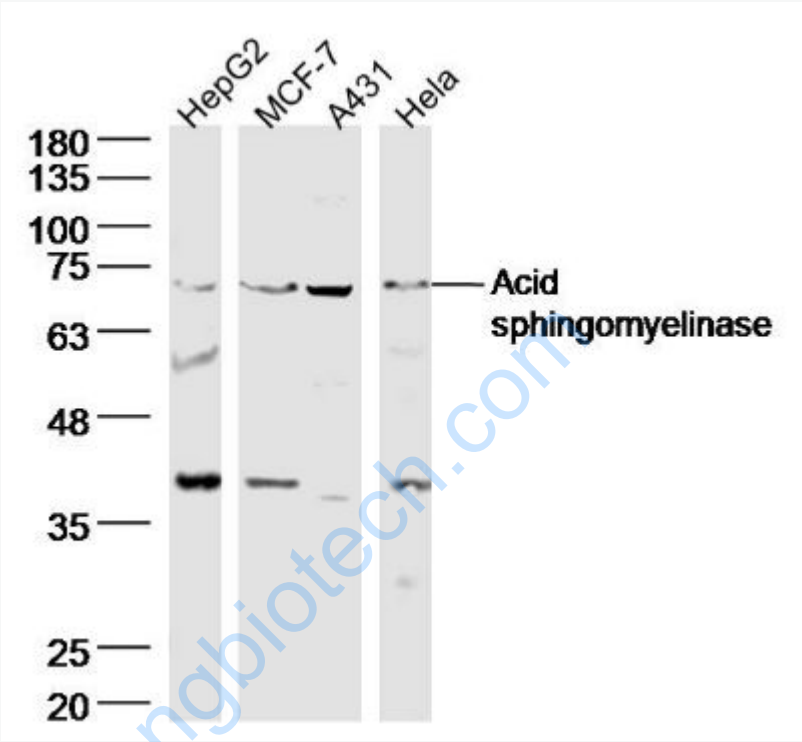
Unigene: 498173Human

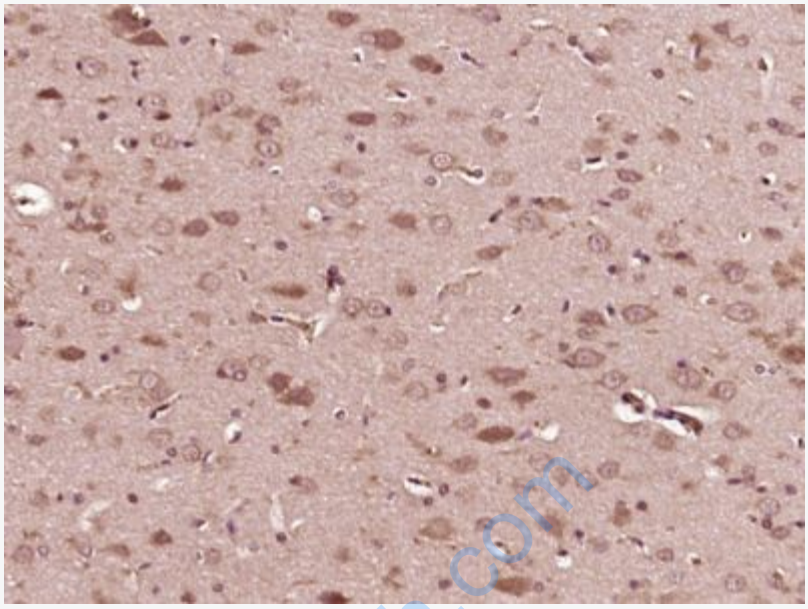
Unigene: 4628Mouse

Unigene: 485064Mouse

Unigene: 18277Rat

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

	ASM酸性神经鞘磷脂酶是ASMase神经鞘磷脂酶最重要的一个亚型,是The cell membrane的重要组成成分。ASM在Apoptosis、调节Tumour细胞生长、参与Fas信号系统传递等方面均可发挥重要作用。
Picture:	 Sample: HepG2(human) cell Lysate at 30 ug MCF-7(human) cell Lysate at 30 ug A431(human) cell Lysate at 30 ug Hale(human) cell Lysate at 30 ug Primary: Anti- Acid sphingomyelinase (SL6318R) at 1/300 dilution Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution Predicted band size: 64kD Observed band size: 69 kD



Paraformaldehyde-fixed, paraffin embedded (Rat brain); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (Acid sphingomyelinase) Polyclonal Antibody, Unconjugated (SL6318R) at 1:400 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.