SunLong Biotech Co.,LTD
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[Primary Antibody]

Rabbit Anti-Acid sphingomyelinase antibody

Catalog Number: SL6318R

FOR RESEARCH USE ONLY

[DATASHEET]

Host:	Rabbit
Isotype:	IgG
Clonality:	Polyclonal
Target:	Acid sphingomyelinase
Immunogen:	KLH conjugated synthetic peptide derived from human Acid sphingomyelinase: 201-300/629
Purification:	affinity purified by Protein A
Concentration:	1mg/ml
Storage:	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol. Shipped at 4℃. Store at -20 °C for one year. Avoid repeated freeze/thaw cycles.
Applications:	WB (1:500-2000) IHC-P (1:100-500) IHC-F (1:100-500) IF (1:100-500) Flow-Cyt (2ug/Test) Not yet tested in other applications. Optimal working dilutions must be determined by the end user.
Reactivity:	Human, Rat (predicted: Mouse, Rabbit, Pig, Cow, Dog)
Predicted MW.:	64kDa
Subcellular Location:	cytoplasmic
Background:	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.

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	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)

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	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. **Database links:** **Entrez Gene:** 505097 Cow **Entrez Gene:** 485334 Dog **Entrez Gene:** 100720041 Guinea pig **Entrez Gene:** 6609 Human **Entrez Gene:** 20597 Mouse **Entrez Gene:** 100353898 Rabbit **Entrez Gene:** 308909 Rat Omim: 607608 Human **SwissProt:** Q0VD19 Cow **SwissProt:** P17405 Human **SwissProt:** Q04519 Mouse **Unigene:** 498173 Human **Unigene:** 4628 Mouse **Unigene:** 485064 Mouse **Unigene:** 18277 Rat ASM acid sphingomyelinase is the most important subtype of ASMase sphingomyelinase and an important component of the cell membrane. ASM plays important roles in apoptosis, regulation of tumour cell growth, and participation in Fas signaling system transmission.

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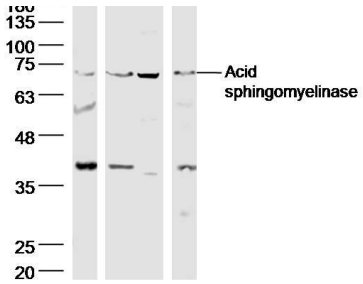
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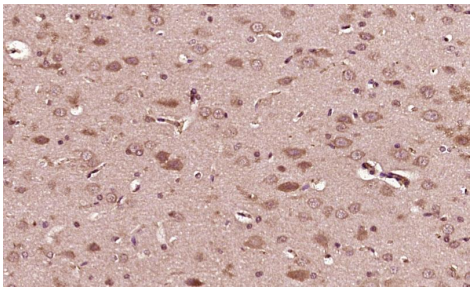
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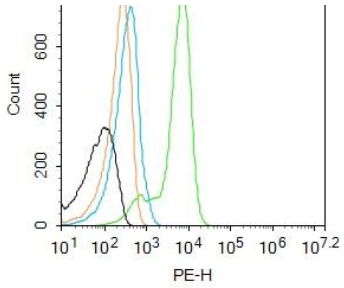
[VALIDATION IMAGES]





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.



Blank control:A431.
Primary Antibody (green line): Rabbit Anti-Acid sphingomyelinase antibody (SL6318R)
Dilution: 2μg /10^6 cells;
Isotype Control Antibody (orange line): Rabbit IgG .
Secondary Antibody : Goat anti-rabbit IgG-AF488
Dilution: 1μg /test.
Protocol
The cells were fixed with 4% PFA (10min at room temperature)and then permeabilized with 0.1% PBST for 20 min at room temperature.The cells were then incubated in 5%BSA to block non-specific protein-protein interactions for 30 min at room temperature .Cells stained with Primary Antibody for 30 min at room temperature. The secondary antibody used for 40 min at room temperature. Acquisition of 20,000 events was performed.

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