# antibodies -online.com





# anti-SMPD1 antibody (AA 201-300) (Cy7)



Go to Product page

$\sim$					
()	VE	۲۱	/1	$\triangle$	Λ

Quantity:	100 μL
Target:	SMPD1
Binding Specificity:	AA 201-300
Reactivity:	Human, Mouse, Rat
Host:	Rabbit
Clonality:	Polyclonal
Conjugate:	This SMPD1 antibody is conjugated to Cy7
Application:	Western Blotting (WB), Immunofluorescence (Cultured Cells) (IF (cc)), Immunofluorescence (Paraffin-embedded Sections) (IF (p))

#### **Product Details**

Immunogen:	KLH conjugated synthetic peptide derived from human Acid sphingomyelinase
Isotype:	IgG
Cross-Reactivity:	Human, Mouse, Rat
Predicted Reactivity:	Dog,Cow,Pig,Rabbit
Purification:	Purified by Protein A.

## **Target Details**

Target:	SMPD1
Alternative Name:	Acid sphingomyelinase (SMPD1 Products)

#### Target Details

Bac	kar	'nΙ	ın	Ч.
Duo		$\sim$	<i>.</i>	ч.

Synonyms: Acid sphingomyelinase, ASM, ASM\_HUMAN, aSMase, NPD, Smpd1, Sphingomyelin phosphodiesterase 1 acid lysosomal, Sphingomyelin phosphodiesterase.

Background: Converts sphingomyelin to ceramide. Also has phospholipase C activities toward 12-diacylglycerolphosphocholine and 12-diacylglycerolphosphoglycerol. Isoform 2 and

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.

Gene ID: 6609

UniProt: P17405

#### **Application Details**

Application Notes: IF(IHC-P) 1:50-200

IF(IHC-F) 1:50-200

IF(ICC) 1:50-200

Restrictions: For Research Use only

#### Handling

Format:	Liquid
Concentration:	1 μg/μL
Buffer:	Aqueous buffered solution containing 0.01M TBS (pH 7.4) with 1 % BSA, 0.03 % Proclin300 and 50 % Glycerol.
Preservative:	ProClin
Precaution of Use:	This product contains ProClin: a POISONOUS AND HAZARDOUS SUBSTANCE, which should be

## Handling

	handled by trained staff only.
Storage:	-20 °C
Storage Comment:	Store at -20°C. Aliquot into multiple vials to avoid repeated freeze-thaw cycles.
Expiry Date:	12 months