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Rabbit Anti-ASAH1 antibody

SL12976R

Product Name:	ASAH1
Chinese Name:	酸性神经酰胺酶1抗体
Alias:	AC; ACDase; Acid CDase; Acid ceramidase; Acid ceramidase precursor; Acid ceramidase subunit beta; Acylsphingosine deacylase; ASAH 1; ASAH; ASAH1; ASAH1_HUMAN; FLJ21558; FLJ22079; N acylsphingosine amidohydrolase (acid ceramidase) 1; N acylsphingosine amidohydrolase 1; N acylsphingosine amidohydrolase; N-acylsphingosine amidohydrolase; PHP; PHP32; Putative 32 kDa heart protein.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Chicken,Dog,Pig,Cow,Horse,Sheep,
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:	29kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human Acid ceramidase subunit beta:301-395/395
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:	Acid ceramidase catalyzes the degradation of ceramide in normal tissues, and deficiency

leads to accumulation of ceramide in tissues, a hallmark of Farber disease. Effected individuals experience early onset joint problems and neurological problems, owing to mutations in the acid ceramidase gene. Bioinformatic analysis of gene expression also reveals acid ceramidase to be among the 5 most important genes associated with melanoma. In addition to ceramide hydrolysis, purified acid ceramidase also exhibits the ability to catalyze ceramide synthesis, utilizing [14C]lauric acid and sphingosine as substrates. Interestingly, pH regulates which reaction is favored; for hydrolysis the pH optimum is 4.5, whereas for the reverse reaction favors a pH of 5.5, further supporting a complex and central role for acid ceramidase in sphingolipid metabolism.

Function:
Hydrolyzes the sphingolipid ceramide into sphingosine and free fatty acid.

Subunit:
Heterodimer of one alpha and one beta subunit.

Subcellular Location:
Lysosome.

Tissue Specificity:
Broadly expressed with highest expression in heart.

DISEASE:
Defects in ASAH1 are the cause of Farber lipogranulomatosis (FL) [MIM:228000]; also known as Farber disease (FD). This sphingolipid disease is characterized by subcutaneous lipid-loaded nodules, excruciating pain in the joints and extremities, marked accumulation of ceramide in lysosomes, and death by three years of age.

Similarity:
Belongs to the acid ceramidase family.

SWISS:
Q13510

Gene ID:
427

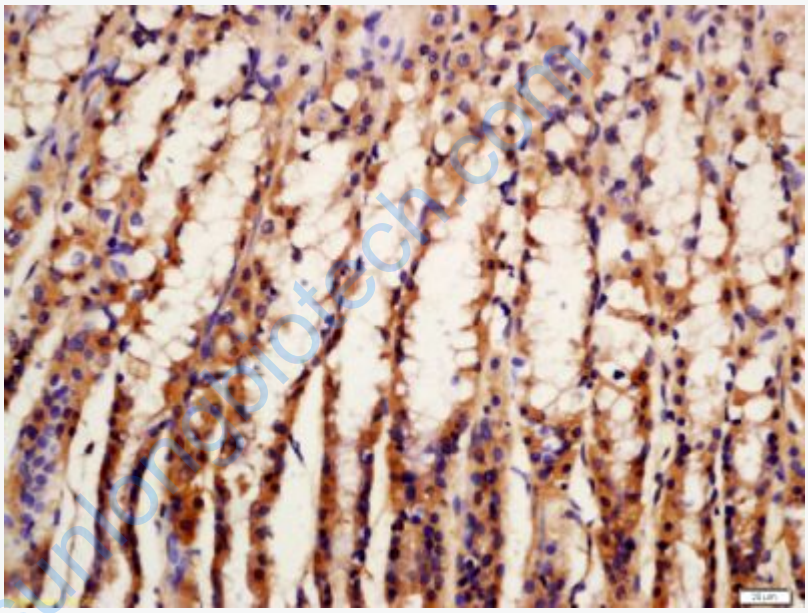
Database links:

Entrez Gene: 427Human

Entrez Gene: 11886Mouse

Entrez Gene: 84431Rat

Omim: 613468Human

	SwissProt: Q13510Human SwissProt: Q9WV54Mouse SwissProt: Q6P7S1Rat **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Picture:	 Tissue/cell: Mouse stomach 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-ASAH1 Polyclonal Antibody, Unconjugated(SL12976R) 1:200, overnight at 4°C, followed by conjugation to the secondary antibody(SP-0023) and DAB(C-0010) staining