

ASAH1 Antibody

Catalog # ASC10740

Product Information

Application	WB, IF, E, IHC-P
Primary Accession	Q13510
Other Accession	EAW63795 , 119584199
Reactivity	Human, Mouse, Rat
Host	Rabbit
Clonality	Polyclonal
Isotype	IgG
Calculated MW	44660
Concentration (mg/ml)	1 mg/mL
Conjugate	Unconjugated
Application Notes	ASAH1 antibody can be used for detection of ASAH1 by Western blot at 1 and 2 μ g/mL. Antibody can also be used for immunohistochemistry starting at 2.5 μ g/mL. For immunofluorescence start at 20 μ g/mL.

Additional Information

Gene ID	427
Other Names	Acid ceramidase, AC, ACDase, Acid CDase, 3.5.1.23, Acylsphingosine deacylase, N-acylsphingosine amidohydrolase, Putative 32 kDa heart protein, PHP32, Acid ceramidase subunit alpha, Acid ceramidase subunit beta, ASAH1, ASAH
Target/Specificity	ASAH1; Multiple isoforms of ASAH1 are known to exist.
Reconstitution & Storage	ASAH1 antibody can be stored at 4°C for three months and -20°C, stable for up to one year. As with all antibodies care should be taken to avoid repeated freeze thaw cycles. Antibodies should not be exposed to prolonged high temperatures.
Precautions	ASAH1 Antibody is for research use only and not for use in diagnostic or therapeutic procedures.

Protein Information

Name	ASAH1 (HGNC:735)
Synonyms	ASAH
Function	Lysosomal ceramidase that hydrolyzes sphingolipid ceramides into sphingosine and free fatty acids at acidic pH (PubMed: 10610716 , PubMed: 11451951 , PubMed: 15655246 , PubMed: 26898341 , PubMed: 36752535 , PubMed: 7744740 , PubMed: 7852294). Ceramides, sphingosine, and its phosphorylated form sphingosine-1-phosphate are

bioactive lipids that mediate cellular signaling pathways regulating several biological processes including cell proliferation, apoptosis and differentiation (PubMed:[10610716](#)). Has a higher catalytic efficiency towards C12- ceramides versus other ceramides (PubMed:[15655246](#), PubMed:[7744740](#)). Also catalyzes the reverse reaction allowing the synthesis of ceramides from fatty acids and sphingosine (PubMed:[12764132](#), PubMed:[12815059](#)). For the reverse synthetic reaction, the natural sphingosine D-erythro isomer is more efficiently utilized as a substrate compared to D- erythro-dihydrosphingosine and D-erythro-phytosphingosine, while the fatty acids with chain lengths of 12 or 14 carbons are the most efficiently used (PubMed:[12764132](#)). Also has an N-acylethanolamine hydrolase activity (PubMed:[15655246](#)). By regulating the levels of ceramides, sphingosine and sphingosine-1-phosphate in the epidermis, mediates the calcium-induced differentiation of epidermal keratinocytes (PubMed:[17713573](#)). Also indirectly regulates tumor necrosis factor/TNF- induced apoptosis (By similarity). By regulating the intracellular balance between ceramides and sphingosine, in adrenocortical cells, probably also acts as a regulator of steroidogenesis (PubMed:[22261821](#)).

Cellular Location Lysosome. Secreted. Note=Secretion is extremely low and localization to lysosomes is mannose-6-phosphate receptor-dependent

Tissue Location Broadly expressed with higher expression in heart.

Background

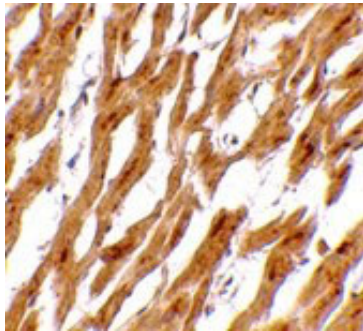
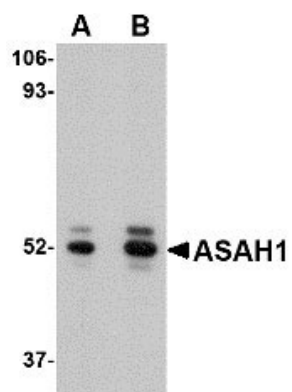
ASAH1 Antibody: Sphingolipids are hydrolyzed by ceramidases to yield sphingosine and fatty acids. These ceramidases are classified according to the pH range that supports their optimal activity. ASAH1 is an acid ceramidase and key regulator of ceramide metabolism. Mutations in this gene results in Farber Lipogranulomatosis, a fatal human genetic disorder that results in the painful swelling of the joints and tendons and pulmonary insufficiency, while a complete knockout of its expression is lethal in mice. Recent studies have shown elevated levels of ASAH1 in Alzheimer's disease (AD) patients correlating with a reduction in sphingomyelin and elevation of ceramide. Pretreatment of cultured neurons with recombinant ASAH1 prevented the cells from undergoing A-beta (Ab)-induced apoptosis.

References

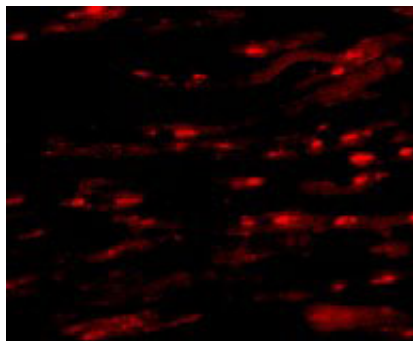
- Nilsson A and Duan RD. Alkaline sphingomyelinases and ceramidases of the gastrointestinal tract. *Chem. Phys. Lipids* 1999; 102:97-105.
- Koch J, Gartner S, Li CM, et al. Molecular cloning and characterization of a full-length complementary DNA encoding human acid ceramidase. Identification of the first molecular lesion causing Farber's disease. *J. Biol. Chem.* 1996; 271:33110-5.
- Li CM, Park JH, Simonaro CM, et al. Insertional mutagenesis of the mouse acid ceramidase gene leads to early embryonic lethality in homozygotes and progressive lipid storage disease in heterozygotes. *Genomics* 2002; 79:218-24.
- He X, Huang Y, Li B, et al. Deregulation of sphingolipid metabolism in Alzheimer's disease. *Neurobiol. Aging* 2010; 31:398-408.

Images

Western blot analysis of ASAH1 in mouse heart tissue lysate with ASAH1 antibody at (A) 1 and (B) 2 µg/mL.



Immunohistochemistry of ASAH1 in human heart tissue with ASAH1 antibody at 2.5 µg/mL.



Immunofluorescence of ASAH1 in Human Heart cells with ASAH1 antibody at 20 µg/mL.

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