

PRODUCT INFORMATION



Acid Ceramidase (human, recombinant)

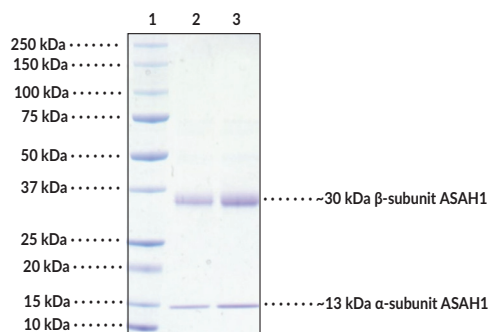
Item No. 31491

Overview and Properties

Synonyms:	AC, ACDase, Acid CDase, Acylsphingosine Deacylase, ASAH, N-Acylethanolamine Hydrolase, N-Acylsphingosine, N-Acylsphingosine Amidohydrolase, PHP32
Source:	Active recombinant human N-terminal His-tagged AC expressed in insect cells
Amino Acids:	22-395
Uniprot No.:	Q13510
Molecular Weight:	30kDa β -subunit, 13 kDa α -subunit
Storage:	-80°C (as supplied)
Stability:	≥ 1 year
Purity:	$\geq 65\%$ estimated by SDS-PAGE
Supplied in:	1x PBS, pH 7.2, 0.02% Triton X-100, 20% glycerol
Protein Concentration:	<i>batch specific</i> mg/ml
Unit Definition:	One unit is defined as the amount of enzyme required to produce 1 nmol of umbelliferone per minute at 37°C in sodium acetate, pH 4.5, buffer with 4 mM RBM 1-151 as the substrate

Information represents the product specifications. Batch specific analytical results are provided on each certificate of analysis.

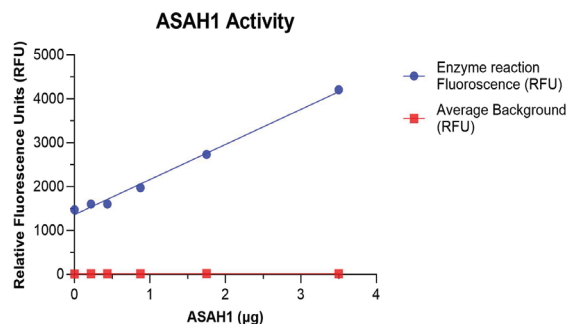
Images



Lane 1: MW Markers
Lane 2: Acid Ceramidase (2 μ g)
Lane 3: Acid Ceramidase (4 μ g)

SDS-PAGE Analysis of Acid Ceramidase.

Representative gel image shown; actual purity may vary between each batch.



ASA1-1 Fluorescence Activity Assay. Acid Ceramidase activity was monitored by fluorescence spectroscopy. An acid ceramidase-specific substrate is cleaved generating a fluorescent signal with λ_{Ex} = 355 nm and λ_{Em} = 460 nm.

WARNING
THIS PRODUCT IS FOR RESEARCH ONLY - NOT FOR HUMAN OR VETERINARY DIAGNOSTIC OR THERAPEUTIC USE.

SAFETY DATA
This material should be considered hazardous until further information becomes available. Do not ingest, inhale, get in eyes, on skin, or on clothing. Wash thoroughly after handling. Before use, the user must review the [complete](#) Safety Data Sheet, which has been sent via email to your institution.

WARRANTY AND LIMITATION OF REMEDY
Buyer agrees to purchase the material subject to Cayman's Terms and Conditions. Complete Terms and Conditions including Warranty and Limitation of Liability information can be found on our website.

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Description

Acid ceramidase (AC) is a hydrolase and member of the N-terminal nucleophile superfamily of hydrolases.^{1,2} It is initially expressed as a proenzyme, which undergoes autocleavage at the peptide bond between threonine 142 and cysteine 143 to produce the active enzyme.³ AC is composed of an α -subunit domain and a β -subunit domain linked by a disulfide bond.^{1,3} It is ubiquitously expressed and located in the lysosome.³ AC is involved in sphingolipid degradation and hydrolyzes ceramides into sphingolipids and free fatty acids.⁴ Mutations in *ASAH1*, the gene encoding AC, are found in patients with Farber disease or spinal muscular atrophy with progressive myoclonic epilepsy (SMA-PME).^{2,5} Cayman's Acid Ceramidase (human, recombinant) protein can be used for enzyme activity assay, ELISA, and Western blot (WB) applications.

References

1. Gebai, A., Gorelik, A., Li, Z., *et al.* Structural basis for the activation of acid ceramidase. *Nat. Commun.* **9**(1), 1621 (2018).
2. Momoi, T., Ben-Yoseph, Y., and Nadler, H.L. Substrate-specificities of acid and alkaline ceramidases in fibroblasts from patients with Farber disease and controls. *Biochem. J.* **205**(2), 419-425 (1982).
3. Ferlinz, K., Kopal, G., Bernardo, K., *et al.* Human acid ceramidase. Processing, glycosylation, and lysosomal targeting. *J. Biol. Chem.* **276**(38), 35352-35360 (2001).
4. Zeidan, Y.H., Jenkins, R.W., Korman, J.B., *et al.* Molecular targeting of acid ceramidase: Implications to cancer therapy. *Curr. Drug Targets* **9**(8), 653-661 (2008).
5. Yu, F.P.S., Amintas, S., Levade, T., *et al.* Acid ceramidase deficiency: Farber disease and SMA-PME. *Orphanet J. Rare Dis.* **13**(1), 121 (2018).

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