abcam

Product datasheet

Recombinant Human Acid sphingomyelinase protein ab132844

1 Abreviews 1 Image

Description

Product name	Recombinant Human Acid sphingomyelinase protein
Expression system	Wheat germ
Protein length	Full length protein
Animal free	No
Nature	Recombinant
Species	Human
Sequence	MPRYGASLRQSCPRSGREQGQDGTAGAPGLLWMGLALA LALALALALSDS RVLWAPAEAHPLSPQGHPARLHRIVPRLRDVFGWGNLTC PICKGLFTAIN LGLKKEPNVARVGSVAIKLCNLLKIAPPAVCRSIVHLFEDD MVEVWRRSV LSPSEACGLLLGSTCGHWDIFSSWNISLPTVPKPPPKPPS PPAPGAPVSR ILFLTDLHWDHDYLEGTDPDCADPLCCRRGSGLPPASRP GAGYWGEYSKC DLPLRTLESLLSGLGPAGPFDMVYWTGDIPAHDVWHQTR QDQLRALTTVT ALVRKFLGPVPVYPAVGNHESTPVNSFPPPFIEGNHSSR WLYEAMAKAWE PWLPAEALRTLRCI
Predicted molecular weight	66 kDa including tags
Amino acids	1 to 364

Specifications

Our **Abpromise guarantee** covers the use of **ab132844** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Applications	Western blot
	SDS-PAGE
	ELISA

Form	Liquid

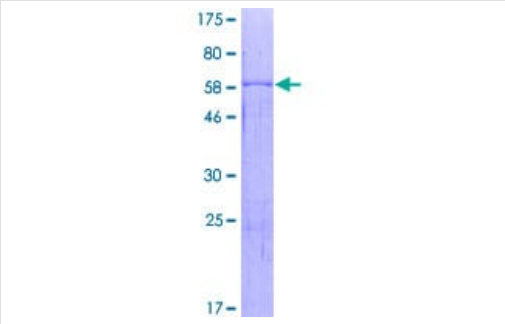
Preparation and Storage

Stability and Storage	Shipped on dry ice. Upon delivery aliquot and store at -80ºC. Avoid freeze / thaw cycles. pH: 8.00 Constituents: 0.31% Glutathione, 0.79% Tris HCl Reduced glutathione

General Info

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.
Involvement in 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) 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.
Sequence similarities	Belongs to the acid sphingomyelinase family. Contains 1 saposin B-type domain.
Cellular localization	Lysosome.

Images



SDS-PAGE - Recombinant Human Acid sphingomyelinase protein (ab132844)

12.5% SDS-PAGE Stained with Coomassie Blue

Please note: All products are "FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC PROCEDURES"

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