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Product datasheet

Recombinant Human PSAP protein ab167924

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Description

Product name	Recombinant Human PSAP protein
Purity	> 95 % Densitometry. ab167924 was purifed using Ni-NTA chromatography.
Endotoxin level	< 1.000 Eu/µg
Expression system	HEK 293 cells
Accession	P07602
Protein length	Full length protein
Animal free	No
Nature	Recombinant
Species	Human
Sequence	ASGPVLGLKE CTRGSAVWCQ NVKTASDCGA VKHCLQTVWN KPTVKSLPCD ICKDVVTAAG DMLKDNATEE EILVYLEKTC DWLPKPNMSA SCKEIVDSYL PVILDIIKGE MSRPGEVCSA LNLCESLQKH LAELNHQKQL ESNKIPELDM TEVVAPFMAN IPLLLYPQDG PRSKPQPKDN GDVCQDCIQM VTDIQTAVRT NSTFVQALVE HVKEECDRLG PGMADICKNY ISQYSEIAIQ MMMHMQPKEI CALVGFCDEV KEMPMQTLVP AKVASKNVIP ALELVEPIKK HEVPAKSDVY CEVCEFLVKE VTKLIDNNKT EKEILDAFDK MCSKLPKSLS EECQEVVDTY GSSILSILLE EVSPELVCSM LHLCSGTRLP ALTVHVTQPK DGGFCEVCKK LVGYLDRNLE KNSTKQEILA ALEKGCSFLP DPYQKQCDQF VAEYEPVLIE ILVEVMDPSF VCLKIGACPS AHKPLLGTEK CIWGPSYWCQ NTETAAQCNA VEHCKRHVWN KLHHHHHH
Predicted molecular weight	58 kDa including tags
Amino acids	17 to 524
Tags	His tag C-Terminus

Specifications

Our **Abpromise guarantee** covers the use of **ab167924** 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 ELISA SDS-PAGE Mass Spectrometry
Mass spectrometry	LC-MS/MS
Form	Lyophilized

Preparation and Storage

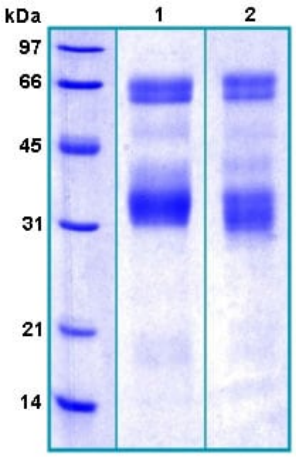
Stability and Storage	Shipped at 4°C. Store at -80°C. Constituents: 99% Phosphate Buffer, 0.43% Sodium chloride
Reconstitution	Add 200µl of deionized water to prepare a working stock solution of 0.5 mg/ml and let the lyophilized pellet dissolve completely. Aliquot reconstituted protein to avoid repeated freezing/thawing cycles and store at -80°C for long term storage. Product is not sterile! Please filter the product by an appropriate sterile filter before using it in the cell culture.

General Info

Function	The lysosomal degradation of sphingolipids takes place by the sequential action of specific hydrolases. Some of these enzymes require specific low-molecular mass, non-enzymic proteins: the sphingolipids activator proteins (coproteins). Saposin-A and saposin-C stimulate the hydrolysis of glucosylceramide by beta-glucosylceramidase (EC 3.2.1.45) and galactosylceramide by beta-galactosylceramidase (EC 3.2.1.46). Saposin-C apparently acts by combining with the enzyme and acidic lipid to form an activated complex, rather than by solubilizing the substrate. Saposin-B stimulates the hydrolysis of galacto-cerebroside sulfate by arylsulfatase A (EC 3.1.6.8), GM1 gangliosides by beta-galactosidase (EC 3.2.1.23) and globotriaosylceramide by alpha-galactosidase A (EC 3.2.1.22). Saposin-B forms a solubilizing complex with the substrates of the sphingolipid hydrolases. Saposin-D is a specific sphingomyelin phosphodiesterase activator (EC 3.1.4.12).
Involvement in disease	Defects in PSAP are the cause of combined saposin deficiency (CSAPD) [MIM:611721]; also known as prosaposin deficiency. CSAPD is due to absence of all saposins, leading to a fatal storage disorder with hepatosplenomegaly and severe neurological involvement. Defects in PSAP saposin-B region are the cause of leukodystrophy metachromatic due to saposin-B deficiency (MLD-SAPB) [MIM:249900]. MLD-SAPB is an atypical form of metachromatic leukodystrophy. It is characterized by tissue accumulation of cerebroside-3-sulfate, demyelination, periventricular white matter abnormalities, peripheral neuropathy. Additional neurological features include dysarthria, ataxic gait, psychomotr regression, seizures, cognitive decline and spastic quadriparesis. Defects in PSAP saposin-C region are the cause of atypical Gaucher disease (AGD) [MIM:610539]. Affected individuals have marked glucosylceramide accumulation in the spleen without having a deficiency of glucosylceramide-beta glucosidase characteristic of classic Gaucher disease, a lysosomal storage disorder. Defects in PSAP saposin-A region are the cause of atypical Krabbe disease (AKRD)

	[MIM:611722]. AKRD is a disorder of galactosylceramide metabolism. AKRD features include progressive encephalopathy and abnormal myelination in the cerebral white matter resembling Krabbe disease. Note=Defects in PSAP saposin-D region are found in a variant of Tay-Sachs disease (GM2-gangliosidosis).
Sequence similarities	Contains 2 saposin A-type domains. Contains 4 saposin B-type domains.
Post-translational modifications	This precursor is proteolytically processed to 4 small peptides, which are similar to each other and are sphingolipid hydrolase activator proteins. N-linked glycans show a high degree of microheterogeneity. The one residue extended Saposin-B-Val is only found in 5% of the chains.
Cellular localization	Lysosome.

Images



SDS-PAGE - Recombinant Human PSAP protein (ab167924)

12% SDS-PAGE analysis of ab167924

Lane 1: reduced and boiled sample, 2.5µg/lane

Lane 2: non-reduced and non-boiled sample, 2.5µg/lane

The ~66 kDa band corresponds to whole PSAP (prosaposin) molecule that consists of four saposin units. The individual saposins naturally cleave off the prosaposin which results in ~15, ~35 and ~50 kDa fragments of mono-, di- and trisaposins, respectively.

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