abcam

Product datasheet

PSAP peptide ab106466

概述

产品名称 PSAP多肽

描述

性质 Synthetic

技术指标

Our Abpromise guarantee covers the use of **ab106466** in the following tested applications.

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

纯度 70 - 90% by HPLC.

补充说明

- First try to dissolve a small amount of peptide in either water or buffer. The more charged residues on a peptide, the more soluble it is in aqueous solutions.
- If the peptide doesn't dissolve try an organic solvent e.g. DMSO, then dilute using water or buffer.
- Consider that any solvent used must be compatible with your assay. If a peptide does not dissolve and you need to recover it, lyophilise to remove the solvent.
- Gentle warming and sonication can effectively aid peptide solubilisation. If the solution is cloudy or has gelled the peptide may be in suspension rather than solubilised.
- Peptides containing cysteine are easily oxidised, so should be prepared in solution just prior to use.

制备和贮存

稳定性和存储 Shipped at 4°C. Upon delivery aliquot and store at -20°C or -80°C. Avoid repeated freeze / thaw cycles.

Information available upon request.

常规信息

功能 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).
疾病相关	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).
序列相似性	Contains 2 saposin A-type domains. Contains 4 saposin B-type domains.
翻译后修饰	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.
细胞定位	Lysosome.

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Terms and conditions

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