abcam

Product datasheet

Anti-PSAP antibody ab97441

2 图像

概述

产品名称	Anti-PSAP抗体
描述	**兔多克隆抗体**to PSAP
宿主	Rabbit
经测试应用	**适用于:** WB, IHC-P
种属反应性	**与反应:** Human **预测可用于:** Cow, Monkey
免疫原	Recombinant protein fragment corresponding to a region within amino acids 1 and 220 of Human PSAP (NP_002769)
阳性对照	293T, A431, HeLa and Molt-4 cell lines. Paraffin-embedded NCIN87 xenograft.

性能

形式	Liquid
存放说明	Shipped at 4°C. Upon delivery aliquot. Store at -20°C or -80°C. Avoid freeze / thaw cycle.
存储溶液	Preservative: 0.01% Thimerosal (merthiolate) Constituents: 20% Glycerol, 0.1M Tris, 0.1M Glycine, pH 7.0
纯度	Immunogen affinity purified
克隆	**多克隆**
同种型	IgG

应用

Our Abpromise guarantee covers the use of **ab97441** in the following tested applications.

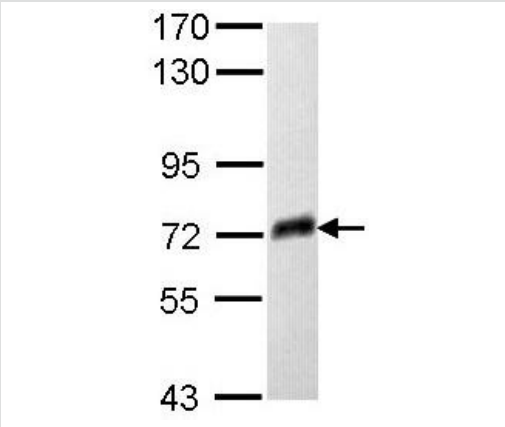
The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

应用	Ab评论	说明
WB		1/500 - 1/3000. Predicted molecular weight: 58 kDa.
IHC-P		1/100 - 1/500.

靶标

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.

图片

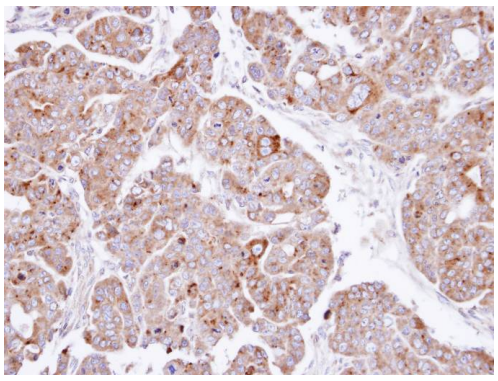


Western blot - Anti-PSAP antibody (ab97441)

Anti-PSAP antibody (ab97441) at 1/2000 dilution + Molt-4 whole cell lysate at 30 µg

Predicted band size: 58 kDa

7.5% SDS PAGE



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-PSAP antibody (ab97441)

Immunohistochemical analysis of paraffin-embedded NCIN87 xenograft using ab97441 at 1/500 dilution.

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