SunLong Biotech Co.,LTD
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[Primary Antibody]

Rabbit Anti-PSAP antibody

Catalog Number: SL2241R

FOR RESEARCH USE ONLY

[DATASHEET]

Host:	Rabbit
Isotype:	IgG
Clonality:	Polyclonal
Target:	PSAP
Immunogen:	KLH conjugated synthetic peptide derived from human Prosaposin: 421-524/524
Purification:	affinity purified by Protein A
Concentration:	1mg/ml
Storage:	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol. Shipped at 4℃. Store at -20 °C for one year. Avoid repeated freeze/thaw cycles.
Applications:	IHC-P=1:100-500 IHC-F=1:100-500 IF=1:100-500 （Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Reactivity:	Human
Predicted MW.:	58kDa
Subcellular Location:	cytoplasmic
Background:	This gene encodes a highly conserved glycoprotein which is a precursor for 4 cleavage products: saposins A, B, C, and D. Each domain of the precursor protein is approximately 80 amino acid residues long with nearly identical placement of cysteine residues and glycosylation sites. Saposins A-D localize primarily to the lysosomal compartment where they facilitate the catabolism of glycosphingolipids with short oligosaccharide groups. The precursor protein exists both as a secretory protein and as an integral membrane protein and has neurotrophic activities. Mutations in this gene have been associated with Gaucher disease, Tay-Sachs disease, and metachromatic leukodystrophy. Alternative splicing results in multiple transcript variants encoding different isoforms. [provided by RefSeq, Jul 2008] **Function:** The lysosomal degradation of sphingolipids takes place by the

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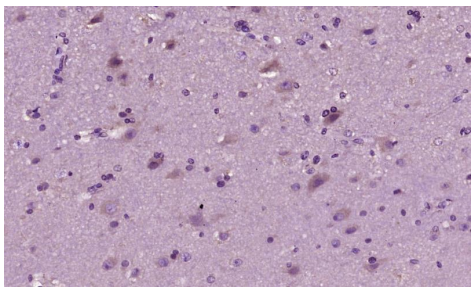
	sequential action of specific hydrolases. Some of these enzymes require specific low-molecular mass, non-enzymic proteins: the sphingolipids activator proteins (coproteins). Saposin-A and saposin-C stimulate the hydrolysis of glucosylceramide by beta-glucosylceramidase (EC 3.2.1.45) and galactosylceramide 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). **Subunit:** Saposin-B is a homodimer. **Subcellular Location:** Lysosome. **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. 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

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	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). **Similarity:** Contains 2 saposin A-type domains. Contains 4 saposin B-type domains. **Database links:** **Entrez Gene:** 5660 Human **Entrez Gene:** 19156 Mouse **Entrez Gene:** 25524 Rat Omim: 176801 Human **SwissProt:** P07602 Human **SwissProt:** Q61207 Mouse **SwissProt:** P10960 Rat **Unigene:** 523004 Human **Unigene:** 277498 Mouse **Unigene:** 97173 Rat

[VALIDATION IMAGES]



Paraformaldehyde-fixed, paraffin embedded (Human brain glioma); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (PSAP) Polyclonal Antibody, Unconjugated (SL2241R) at 1:400 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.

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