



Rabbit Anti-PSAP antibody

SL1879R

Product Name:	PSAP
Chinese Name:	鞘脂激活蛋白原抗体
Alias:	Prosaposin; A1 activator; Cerebroside sulfate activator; Co-beta-glucosidase; Component C; CSAct; Dispersin; GLBA; Glucosylceramidase activator; Proactivator polypeptide; Proactivator polypeptide precursor; Prosaposin (sphingolipid activator protein 1); prosaposin (variant Gaucher disease and variant metachromatic leukodystrophy); Protein A; Protein C; PSAP; SAP-1; SAP-2; SAP_HUMAN; SAP1; Saposin A; Saposin B; Saposin B Val; Saposin C; Saposin D; Saposin-D; Saposins; Sgp1; Sphingolipid activator protein 1; Sphingolipid activator protein 2; Sulfated glycoprotein 1; Sulfatide/GM1 activator.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800IF=1:100-500 (Paraffin sections need antigen repair) not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	8.8/58kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human Prosaposin:101-200/524
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
PubMed:	PubMed

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 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,

Product Detail:

peripheral neuropathy. Additional neurological features include dysarthria, ataxic gait, psychomotor 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).

Similarity:

Contains 2 saposin A-type domains.

Contains 4 saposin B-type domains.

SWISS:

P07602

Gene ID:

5660

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

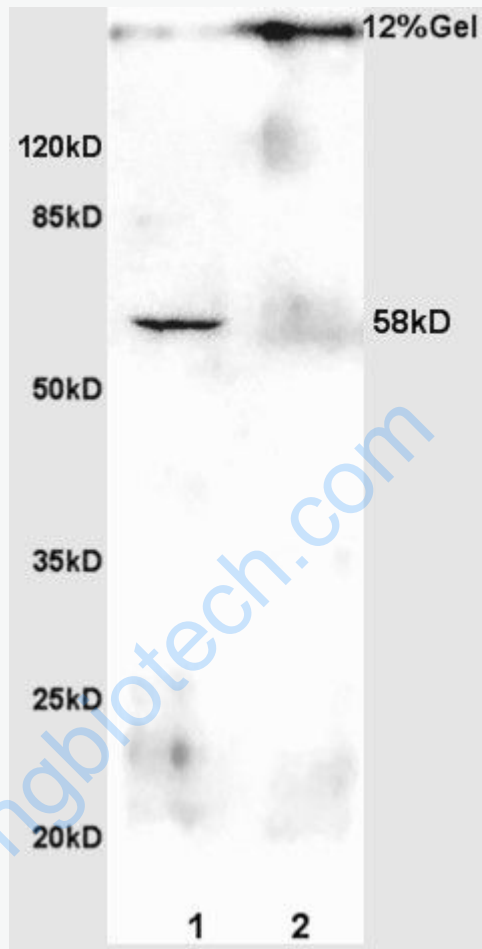
[Unigene: 277498](#)Mouse

[Unigene: 97173](#)Rat

Important Note:

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

表达在正常的前列腺、前列腺增生和前列腺Tumour中。



Picture:

Protein:

testis(rat) lysates at 30ug;

brain(rat) lysates at 30ug;

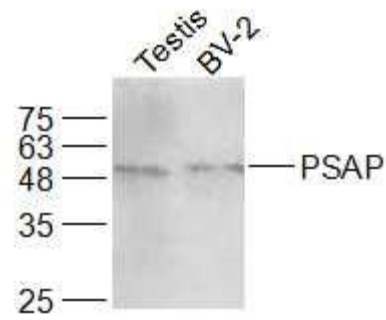
Primary: Anti-PSAP/PAP (SL1879R) at 1:200;

Secondary: HRP conjugated Goat Anti-Rabbit IgG(SL1879R) at 1: 3000;

ECL excited the fluorescence;

Predicted band size : 58kD

Observed band size : 58kD



Sample:

Testis (Rat) Lysate at 40 ug

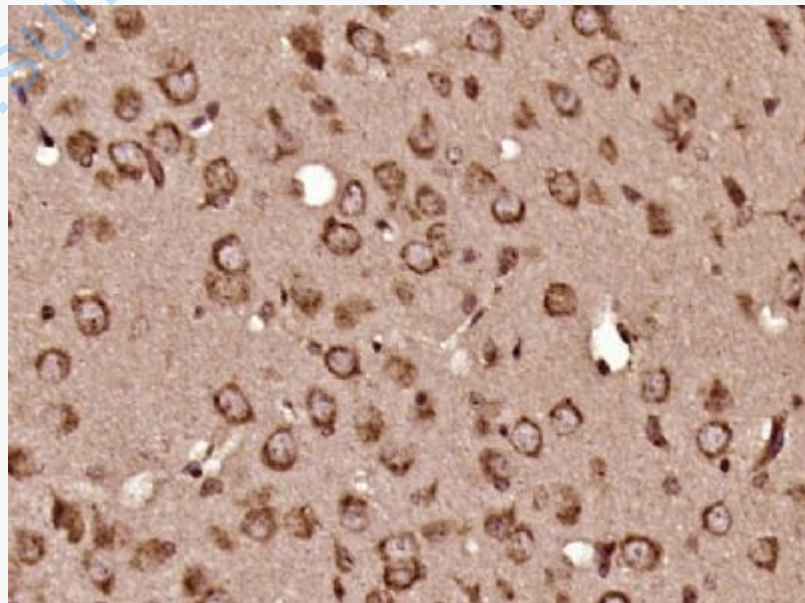
BV-2 (Mouse) CellLysate at 30 ug

Primary: Anti-PSAP (SL1879R) at 1/1000 dilution

Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution

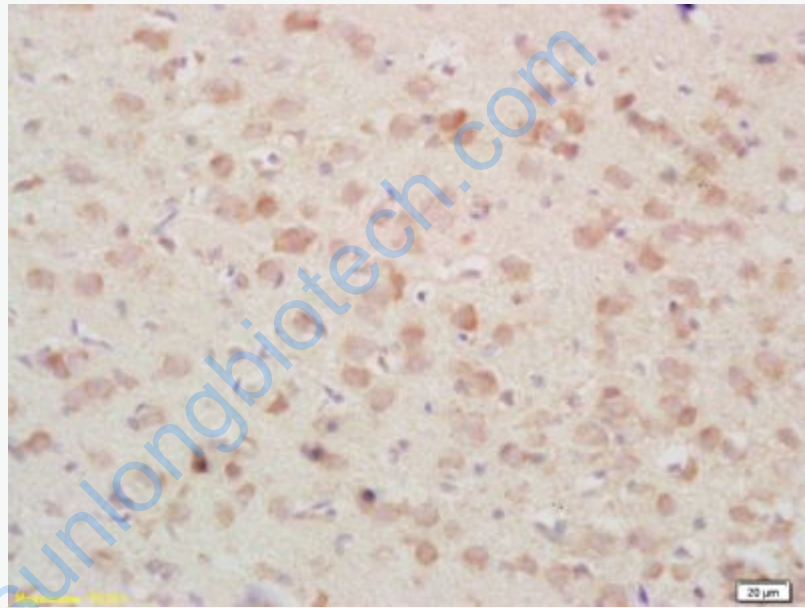
Predicted band size: 8.8/58 kD

Observed band size: 58 kD



Paraformaldehyde-fixed, paraffin embedded (Mouse brain); 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 (SL1879R) at 1:400 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructions and DAB staining.



Tissue/cell: rat brain tissue; 4% Paraformaldehyde-fixed and paraffin-embedded;
Antigen retrieval: citrate buffer (0.01M, pH 6.0), Boiling bathing for 15min; Block endogenous peroxidase by 3% Hydrogen peroxide for 30min; Blocking buffer (normal goat serum,C-0005) at 37°C for 20 min;
Incubation: Anti-PSAP/PAP Polyclonal Antibody, Unconjugated(SL1879R) 1:200, overnight at 4°C, followed by conjugation to the secondary antibody(SP-0023) and DAB(C-0010) staining