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Rabbit Anti-PSAP antibody

SL2241R

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,
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:	58kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human Prosaposin:421-524/524
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

Product Detail:	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,

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).

Similarity:
Contains 2 saposin A-type domains.
Contains 4 saposin B-type domains.

SWISS:
P07602

Gene ID:
5660

Database links:

Entrez Gene: 5660Human

Entrez Gene: 19156Mouse

Entrez Gene: 25524Rat

Omim: 176801Human

SwissProt: P07602Human

SwissProt: Q61207Mouse

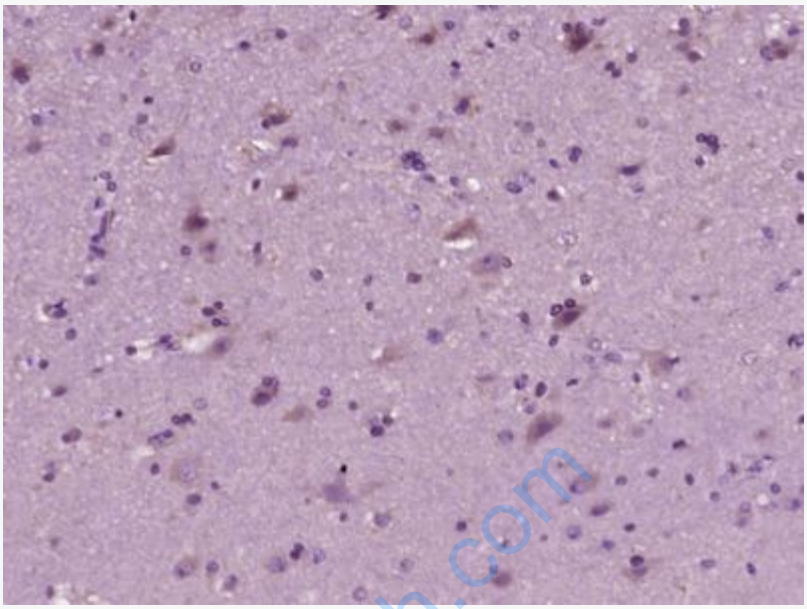
SwissProt: P10960Rat

Unigene: 523004Human

Unigene: 277498Mouse

Unigene: 97173Rat

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

Picture:	 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.