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Rabbit Anti-Protective protein/Cathepsin A antibody

SL6040R

Product Name:	Protective protein/Cathepsin A
Chinese Name:	组织蛋白酶A抗体
Alias:	Cathepsin A; CTSA; BETA GALACTOSIDASE PROTECTIVE PROTEIN; Carboxypeptidase C; Glactosialidosis; GLB2; Goldberg Syndrome; GSL; Lysosomal protective protein; NEURAMINIDASE BETA GALACTOSIDASE EXPRESSION; NGBE; NGBE; PPCA; PPGB; Protective protein for beta galactosidase; PPGB_HUMAN.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Pig,Cow,Horse,
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:	51kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human Protective protein:401-480/480
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:	Lysosomal protective protein/cathepsin A (PPCA) is a lysosomal serine

carboxypeptidase that forms an intralysosomal enzyme complex with ?galactosidase and neuraminidase (NEU1). PPCA is synthesized as a 54 kDa precursor/zymogen, and proteolytically cleaved in the lysosome into a catalytically active 32 and 20 kDa two chain enzyme. The enzyme has cathepsin A activity at acidic pH but maintains also a deamidase/esterase activity at neutral pH. Furthermore, the human enzyme, purified from platelets and lymphocytes, has been shown to function on the inactivation of selected neuropeptides, like substance P, oxytocin, and endothelin I. The autosomal recessive genetic deficiency of PPCA causes galactosialidosis, a neurodegenerative lysosomal storage disorder, resulting in the secondary deficiencies of ?galactosidase and NEU1.

Function:
Protective protein appears to be essential for both the activity of beta-galactosidase and neuraminidase, it associates with these enzymes and exerts a protective function necessary for their stability and activity. This protein is also a carboxypeptidase and can deamidate tachykinins.

Subunit:
Heterodimer of a 32 kDa chain and a 20 kDa chain; disulfide-linked.

Subcellular Location:
Lysosome.

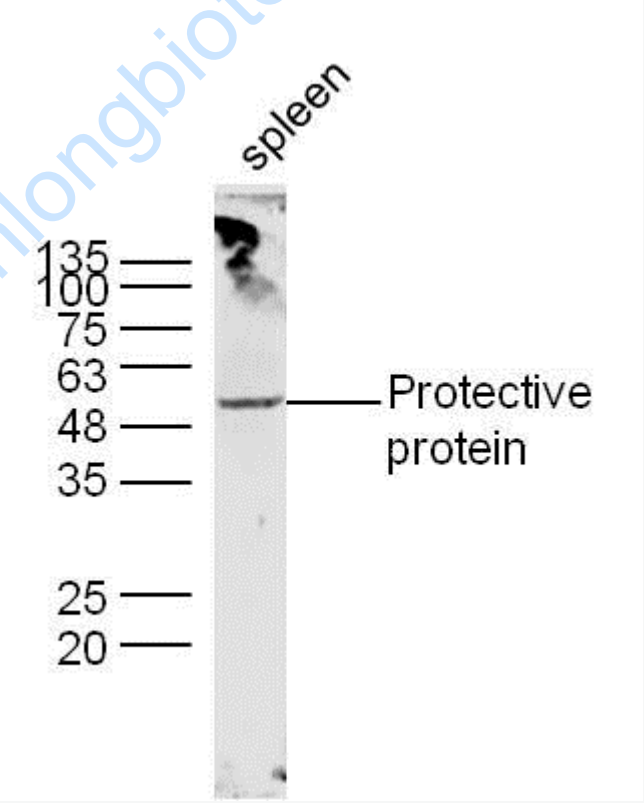
DISEASE:
Defects in CTSA are the cause of galactosialidosis (GSL) [MIM:256540]. A lysosomal storage disease associated with a combined deficiency of beta-galactosidase and neuraminidase, secondary to a defect in cathepsin A. All patients have clinical manifestations typical of a lysosomal disorder, such as coarse facies, cherry red spots, vertebral changes, foam cells in the bone marrow, and vacuolated lymphocytes. Three phenotypic subtypes are recognized. The early infantile form is associated with fetal hydrops, edema, ascites, visceromegaly, skeletal dysplasia, and early death. The late infantile type is characterized by hepatosplenomegaly, growth retardation, cardiac involvement, and a normal or mildly affected mental state. The juvenile/adult form is characterized by myoclonus, ataxia, angiokeratoma, mental retardation, neurologic deterioration, absence of visceromegaly, and long survival.

Similarity:
Belongs to the peptidase S10 family.

SWISS:
P10619

Gene ID:
5476

Database links:

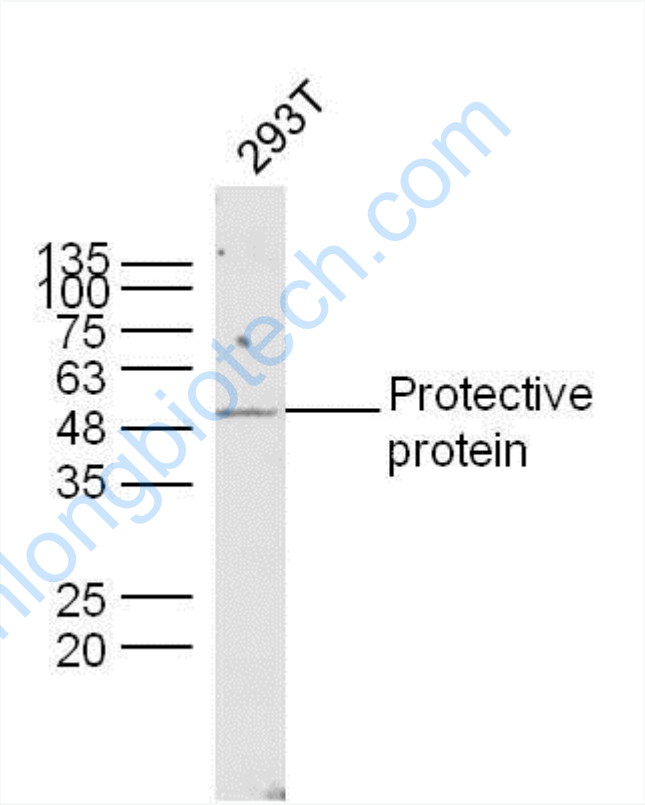
	Entrez Gene: 5476Human Entrez Gene: 19025Mouse Omim: 613111Human SwissProt: P10619Human SwissProt: P16675Mouse Unigene: 609336Human Unigene: 359633Mouse Unigene: 474586Mouse **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Picture:	 Sample: Spleen (Mouse) Lysate at 40 ug

Primary: Anti-Protective protein (SL6040R) at 1/300 dilution

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

Predicted band size: 51 kD

Observed band size: 51 kD



Sample:

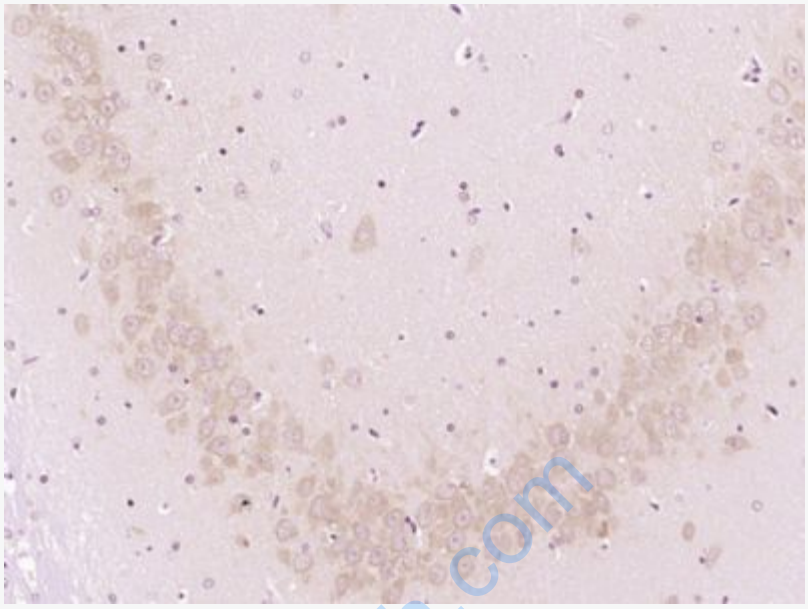
293T Cell (Human) Lysate at 30 ug

Primary: Anti-Protective protein (SL6040R) at 1/300 dilution

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

Predicted band size: 51 kD

Observed band size: 51 kD



Paraformaldehyde-fixed, paraffin embedded (Rat 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 (Protective protein,Cathepsin A) Polyclonal Antibody, Unconjugated (SL6040R) at 1:400 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.