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Rabbit Anti-TPP1 antibody

SL8528R

Product Name:	TPP1
Chinese Name:	细胞生长抑制基因1蛋白抗体
Alias:	Cell growth inhibiting gene 1 protein; Cell growth-inhibiting gene 1 protein; Ceroid lipofuscinosis neuronal 2; Ceroid lipofuscinosis neuronal 2 late infantile (Jansky Bielschowsky disease); Ceroid lipofuscinosis neuronal 2 late infantile antibody CLN 2; CLN2; CLN-2; GIG 1; GIG1; Growth inhibiting protein 1; LPIC; Lysosomal pepstatin insensitive protease; Lysosomal pepstatin-insensitive protease; TPP 1; TPP I; TPP-1; TPP-I; Tpp1; TPP1_HUMAN; TPPI; Tripeptidyl aminopeptidase; Tripeptidyl peptidase I; Tripeptidyl-peptidase 1; Tripeptidyl-peptidase I.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Pig,Cow,Horse,Rabbit,Sheep,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800IF=1:50-200（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	40kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human TPP1/CLN2:401-500/563
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 serine protease with tripeptidyl-peptidase I activity. May act as a non-

specific lysosomal peptidase which generates tripeptides from the breakdown products produced by lysosomal proteinases. Requires substrates with an unsubstituted N-terminus.

Function:
Lysosomal serine protease with tripeptidyl-peptidase I activity. May act as a non-specific lysosomal peptidase which generates tripeptides from the breakdown products produced by lysosomal proteinases. Requires substrates with an unsubstituted N-terminus (By similarity).

Subunit:
Monomer.

Subcellular Location:
Lysosome. Melanosome. Identified by mass spectrometry in melanosome fractions from stage I to stage IV.

Tissue Specificity:
Detected in all tissues examined with highest levels in heart and placenta and relatively similar levels in other tissues.

Post-translational modifications:
Activated by autocatalytic proteolytical processing upon acidification. N-glycosylation is required for processing and activity.

DISEASE:
Involvement in disease: Defects in TPP1 are the cause of neuronal ceroid lipofuscinosis type 2 (CLN2) . A form of neuronal ceroid lipofuscinosis. Neuronal ceroid lipofuscinoses are progressive neurodegenerative, lysosomal storage diseases characterized by intracellular accumulation of autofluorescent liposomal material, and clinically by seizures, dementia, visual loss, and/or cerebral atrophy. The lipopigment pattern seen most often in CLN2 consists of curvilinear profiles.

Similarity:
Belongs to the peptidase S53 family.

SWISS:
O14773

Gene ID:
1200

Database links:

Entrez Gene: 1200Human

	Omim: 607998Human SwissProt: O14773Human Unigene: 523454Human **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.