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TPP1 Recombinant Rabbit mAb

Catalog # AP100068

Product Information

Primary Accession	O14773
Other Accession	O14773
Reactivity	Human
Host	Rabbit
Clonality	Recombinant
Calculated MW	61248
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human TPP1
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.22 □m filtered solution in 20mM hepes7.4, 150mm NaCl, 50% glycerin,0.03% proclin.
SIMILARITY	Belongs to the peptidase S53 family.
SUBUNIT	Monomer.
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.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	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.

Additional Information

Gene ID	1200
Other Names	CLN2; GIG1; LPIC; SCAR7; TPP-1; TPP1_HUMAN; TPP1; Cell growth-inhibiting gene 1 protein; Lysosomal pepstatin-insensitive protease (LPIC); Tripeptidyl aminopeptidase; Tripeptidyl-peptidase I (TPP-I); 3.4.14.9; tripeptidyl peptidase 1; ceroid-lipofuscinosis, neuronal 2, late infantile (Jansky-Bielschowsky disease); spinocerebellar ataxia, autosomal recessive 7; tripeptidyl peptidase I; TPP I; lysosomal pepstatin-insensitive carboxypeptidase
Dilution	ICC/IF=1:50-200

Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. 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.

Protein Information

Name	TPP1
Synonyms	CLN2
Function	Lysosomal serine protease with tripeptidyl-peptidase I activity (PubMed:11054422, PubMed:19038966, PubMed:19038967). May act as a non-specific lysosomal peptidase which generates tripeptides from the breakdown products produced by lysosomal proteinases (PubMed:11054422, PubMed:19038966, PubMed:19038967). Requires substrates with an unsubstituted N-terminus (PubMed:19038966).
Cellular Location	Lysosome. Melanosome. Note=Identified by mass spectrometry in melanosome fractions from stage I to stage IV
Tissue Location	Detected in all tissues examined with highest levels in heart and placenta and relatively similar levels in other tissues

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.