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TPP1 Antibody (N-term)

Affinity Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP17047a

Product Information

Application	WB, E
Primary Accession	O14773
Other Accession	Q60HH1, NP_000382.3
Reactivity	Human
Predicted	Monkey
Host	Rabbit
Clonality	Polyclonal
Isotype	Rabbit IgG
Clone Names	RB36843
Calculated MW	61248
Antigen Region	6-34

Additional Information

Gene ID	1200
Other Names	Tripeptidyl-peptidase 1, TPP-1, Cell growth-inhibiting gene 1 protein, Lysosomal pepstatin-insensitive protease, LPIC, Tripeptidyl aminopeptidase, Tripeptidyl-peptidase I, TPP-I, TPP1, CLN2
Target/Specificity	This TPP1 antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide between 6-34 amino acids from the N-terminal region of human TPP1.
Dilution	WB~~1:1000 E~~Use at an assay dependent concentration.
Format	Purified polyclonal antibody supplied in PBS with 0.09% (W/V) sodium azide. This antibody is purified through a protein A column, followed by peptide affinity purification.
Storage	Maintain refrigerated at 2-8°C for up to 2 weeks. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.
Precautions	TPP1 Antibody (N-term) is for research use only and not for use in diagnostic or therapeutic procedures.

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

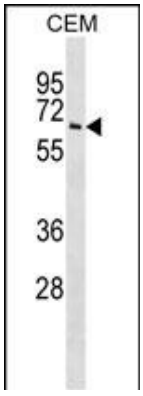
Background

This gene encodes a member of the sedolisin family of serine proteases. The protease functions in the lysosome to cleave N-terminal tripeptides from substrates, and has weaker endopeptidase activity. It is synthesized as a catalytically-inactive enzyme which is activated and auto-proteolyzed upon acidification. Mutations in this gene result in late-infantile neuronal ceroid lipofuscinosis, which is associated with the failure to degrade specific neuropeptides and a subunit of ATP synthase in the lysosome.

References

Souweidane, M.M., et al. J Neurosurg Pediatr 6(2):115-122(2010)
Walus, M., et al. Hum. Mutat. 31(6):710-721(2010)
Latrick, C.M., et al. EMBO J. 29(5):924-933(2010)
Kuizon, S., et al. PLoS ONE 5 (8), E11929 (2010) :
Goldberg-Stern, H., et al. Pediatr. Neurol. 41(4):297-300(2009)

Images



TPP1 Antibody (N-term) (Cat. #AP17047a) western blot analysis in CEM cell line lysates (35ug/lane).This demonstrates the TPP1 antibody detected the TPP1 protein (arrow).