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Immunotag™ TPP1 Polyclonal Antibody

Antibody Specification

Catalog No.	ITN2334
Product Description	Immunotag™ TPP1 Polyclonal Antibody
Size	50 µg, 100 µg
Conjugation	HRP, Biotin, FITC, Alexa Fluor® 350, Alexa Fluor® 405, Alexa Fluor® 488, Alexa Fluor® 555, Alexa Fluor® 594, Alexa Fluor® 647
IMPORTANT NOTE	This product is custom manufactured with a lead time of 3-4 weeks. Once in production, this item cannot be cancelled from an order and is not eligible for return.
Target Protein	TPP1
Clonality	Polyclonal
Storage/Stability	-20°C/1 year
Application	WB,ELISA
Recommended Dilution	WB 1:500-2000 ELISA 1:5000-20000
Concentration	1 mg/ml
Reactive Species	Human,Rat,Mouse
Host Species	Rabbit
Immunogen	Synthesized peptide derived from human protein, at AA range: 10-90
Specificity	TPP1 Polyclonal Antibody detects endogenous levels of protein.
Purification	The antibody was affinity-purified from rabbit antiserum by affinity-chromatography using epitope-specific immunogen
Form	Liquid in PBS containing 50% glycerol, and 0.02% sodium azide.
Gene Name	TPP1 CLN2 GIG1 UNQ267/PRO304
Accession No.	O14773 O89023 Q9EQV6

Antibody Specification

Description	tripeptidyl peptidase 1(TPP1) Homo sapiens 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. [provided by RefSeq, Jul 2008],
Cell Pathway/ Category	Lysosome,
Protein Expression	Adipose tissue,Brain cortex,Liver,Lymph,Placenta,
Subcellular Localization	mitochondrion,lysosome,melanosome,lysosomal lumen,extracellular exosome,
Protein Function	catalytic activity:Release of an N-terminal tripeptide from a polypeptide, but also has endopeptidase activity.,caution:Ref.3 sequence is wrongly reported to originate from bovine.,disease:Defects in TPP1 are the cause of classical late-infantile neuronal ceroid lipofuscinosis (LINCL) [MIM:204500]; also known as ceroid lipofuscinosis neuronal 2 (CLN2). LINCL is a fatal childhood neurodegenerative disease characterized by progressive visual and mental decline, motor disturbance, epilepsy and behavioral changes. The three main subtypes of childhood NCLs defined by the age of onset, clinical features, and ultrastructural morphology are infantile NCL (INCL), classical late-infantile NCL (LINCL), or juvenile NCL (JNCL), although a number of other distinct variants forms have been described.,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.,online information:Neural Ceroid Lipofuscinoses mutation db,PTM:Activated by autocatalytic proteolytical processing upon acidification.,similarity:Belongs to the peptidase S53 family.,subcellular location: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.,
Usage	For Research Use Only! Not for diagnostic or therapeutic procedures.