

Goat Anti-CLN2 / TPP1 Antibody

Peptide-affinity purified goat antibody Catalog # AF1253a

Specification

Goat Anti-CLN2 / TPP1 Antibody - Product Information

Application Primary Accession Other Accession Reactivity Predicted Host Clonality Concentration Isotype Calculated MW WB <u>O14773</u> <u>NP_000382</u>, <u>1200</u>, <u>12751 (mouse)</u> Human Dog, Cow Goat Polyclonal 100ug/200ul IgG 61248

Goat Anti-CLN2 / TPP1 Antibody - Additional Information

Gene ID 1200

Other Names

Tripeptidyl-peptidase 1, TPP-1, 3.4.14.9, Cell growth-inhibiting gene 1 protein, Lysosomal pepstatin-insensitive protease, LPIC, Tripeptidyl aminopeptidase, Tripeptidyl-peptidase I, TPP-I, TPP1, CLN2

Format

0.5 mg lgG/ml in Tris saline (20mM Tris pH7.3, 150mM NaCl), 0.02% sodium azide, with 0.5% bovine serum albumin

Storage

Maintain refrigerated at 2-8°C for up to 6 months. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.

Precautions Goat Anti-CLN2 / TPP1 Antibody is for research use only and not for use in diagnostic or therapeutic procedures.

Goat Anti-CLN2 / TPP1 Antibody - Protein Information

Name TPP1

Synonyms CLN2

Function

Lysosomal serine protease with tripeptidyl-peptidase I activity (PubMed:11054422, PubMed:<a



href="http://www.uniprot.org/citations/19038966" target="_blank">19038966, PubMed:19038966). May act as a
non-specific lysosomal peptidase which generates tripeptides from the breakdown products
produced by lysosomal proteinases (PubMed:<a href="http://www.uniprot.org/citations/11054422"
target="_blank">11054422, PubMed:<a href="http://www.uniprot.org/citations/19038966"
target="_blank">19038966). Requires substrates with an unsubstituted N-terminus
(PubMed:19038966). 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

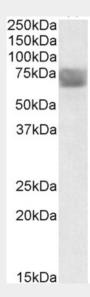
Detected in all tissues examined with highest levels in heart and placenta and relatively similar levels in other tissues

Goat Anti-CLN2 / TPP1 Antibody - Protocols

Provided below are standard protocols that you may find useful for product applications.

- <u>Western Blot</u>
- Blocking Peptides
- Dot Blot
- Immunohistochemistry
- Immunofluorescence
- Immunoprecipitation
- Flow Cytomety
- <u>Cell Culture</u>

Goat Anti-CLN2 / TPP1 Antibody - Images



AF1253a (0.3 μ g/ml) staining of Human Placenta lysate (35 μ g protein in RIPA buffer) with (B) and without (A) blocking with the immunising peptide. Primary incubation was 1 hour. Detected by chemiluminescence.

Goat Anti-CLN2 / TPP1 Antibody - 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.

Goat Anti-CLN2 / TPP1 Antibody - References

Gene therapy for late infantile neuronal ceroid lipofuscinosis: neurosurgical considerations. Souweidane MM, et al. J Neurosurg Pediatr, 2010 Aug. PMID 20672930.

POT1-TPP1 enhances telomerase processivity by slowing primer dissociation and aiding translocation. Latrick CM, et al. EMBO J, 2010 Mar 3. PMID 20094033.

Late infantile neuronal ceroid lipofuscinosis: a new mutation in Arabs. Goldberg-Stern H, et al. Pediatr Neurol, 2009 Oct. PMID 19748052.

Lysosomal serine protease CLN2 regulates tumor necrosis factor-alpha-mediated apoptosis in a Bid-dependent manner. Autefage H, et al. J Biol Chem, 2009 Apr 24. PMID 19246452. Mutations in CLN7/MFSD8 are a common cause of variant late-infantile neuronal ceroid

lipofuscinosis. Kousi M, et al. Brain, 2009 Mar. PMID 19201763.