

# TPP1 Recombinant Rabbit mAb

Catalog # AP100068

## Product Information

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| <b>Primary Accession</b>                | <a href="#">O14773</a>                                                                                                                                                                                                                                                                                                                                                                                                                                                                          |
| <b>Other Accession</b>                  | <a href="#">O14773</a>                                                                                                                                                                                                                                                                                                                                                                                                                                                                          |
| <b>Host</b>                             | Rabbit                                                                                                                                                                                                                                                                                                                                                                                                                                                                                          |
| <b>Clonality</b>                        | Recombinant                                                                                                                                                                                                                                                                                                                                                                                                                                                                                     |
| <b>Calculated MW</b>                    | 61248                                                                                                                                                                                                                                                                                                                                                                                                                                                                                           |
| <b>Physical State</b>                   | Liquid                                                                                                                                                                                                                                                                                                                                                                                                                                                                                          |
| <b>Immunogen</b>                        | KLH conjugated synthetic peptide derived from human TPP1                                                                                                                                                                                                                                                                                                                                                                                                                                        |
| <b>Isotype</b>                          | IgG                                                                                                                                                                                                                                                                                                                                                                                                                                                                                             |
| <b>Purity</b>                           | affinity purified by Protein A                                                                                                                                                                                                                                                                                                                                                                                                                                                                  |
| <b>Buffer</b>                           | 0.22 M filtered solution in 20mM hepes7.4, 150mm NaCl, 50% glycerin,0.03% proclin.                                                                                                                                                                                                                                                                                                                                                                                                              |
| <b>SIMILARITY</b>                       | Belongs to the peptidase S53 family.                                                                                                                                                                                                                                                                                                                                                                                                                                                            |
| <b>SUBUNIT</b>                          | Monomer.                                                                                                                                                                                                                                                                                                                                                                                                                                                                                        |
| <b>Post-translational modifications</b> | Activated by autocatalytic proteolytical processing upon acidification. N-glycosylation is required for processing and activity.                                                                                                                                                                                                                                                                                                                                                                |
| <b>DISEASE</b>                          | 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. |
| <b>Important Note</b>                   | This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.                                                                                                                                                                                                                                                                                                                                                                       |
| <b>Background Descriptions</b>          | 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

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| <b>Gene ID</b>     | 1200                                                                                                                                                                                                                                                                                                                                                                                                                                                       |
| <b>Other Names</b> | 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 |
| <b>Dilution</b>    | ICC/IF=1:50-200                                                                                                                                                                                                                                                                                                                                                                                                                                            |
| <b>Storage</b>     | 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

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| <b>Name</b>              | TPP1                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                    |
| <b>Synonyms</b>          | CLN2                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                    |
| <b>Function</b>          | Lysosomal serine protease with tripeptidyl-peptidase I activity (PubMed: <a href="#">11054422</a> , PubMed: <a href="#">19038966</a> , PubMed: <a href="#">19038967</a> ). May act as a non-specific lysosomal peptidase which generates tripeptides from the breakdown products produced by lysosomal proteinases (PubMed: <a href="#">11054422</a> , PubMed: <a href="#">19038966</a> , PubMed: <a href="#">19038967</a> ). Requires substrates with an unsubstituted N-terminus (PubMed: <a href="#">19038966</a> ). |
| <b>Cellular Location</b> | Lysosome. Melanosome. Note=Identified by mass spectrometry in melanosome fractions from stage I to stage IV                                                                                                                                                                                                                                                                                                                                                                                                             |
| <b>Tissue Location</b>   | 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'.