

CLN2 (L-12): sc-34867

BACKGROUND

CLN2, also known as Tripeptidyl peptidase I (TTP-I), a member of the family of serine-carboxyl proteinases (S53), plays a crucial role in lysosomal protein degradation, and a deficiency in this enzyme leads to fatal neurodegenerative disease. CLN2 is a lysosomal aminopeptidase that sequentially removes tripeptides from small polypeptides and also shows a minor endoprotease activity. In lysosomes, CLN2 proenzyme is converted into a mature enzyme with the assistance of another protease and is able to autoactivate in acidic pH *in vitro* via an unimolecular mechanism.

REFERENCES

1. Golabek, A.A., et al. 2004. Maturation of human tripeptidyl peptidase I *in vitro*. *J. Biol. Chem.* 279: 31058-31067.
2. Golabek, A.A., et al. 2005. Glycosaminoglycans modulate activation, activity and stability of tripeptidyl peptidase I *in vitro* and *in vivo*. *J. Biol. Chem.* 280: 7550-7561.
3. Kohan, R., et al. 2005. Palmitoyl protein thioesterase1 (PPT1) and tripeptidyl peptidase I (TPP-I) are expressed in the human saliva. A reliable and non-invasive source for the diagnosis of infantile (CLN1) and late infantile (CLN2) neuronal ceroid lipofuscinoses. *Clin. Biochem.* 38: 492-494.
4. Oyama, H., et al. 2005. Catalytic residues and substrate specificity of recombinant human tripeptidyl peptidase I (CLN2). *J. Biochem.* 138: 127-134.
5. Sondhi, D., et al. 2005. AAV2-mediated CLN2 gene transfer to rodent and non-human primate brain results in long-term TPP-I expression compatible with therapy for LINCL. *Gene Ther.* 12: 1618-1632.
6. Walus, M., et al. 2005. Ser 475, Glu 272, Asp 276, Asp 327 and Asp 360 are involved in catalytic activity of human tripeptidyl peptidase I. *FEBS Lett.* 579: 1383-1388.

CHROMOSOMAL LOCATION

Genetic locus: TPP1 (human) mapping to 11p15.4; Tpp1 (mouse) mapping to 7 E3.

SOURCE

CLN2 (L-12) is an affinity purified goat polyclonal antibody raised against a peptide mapping at the N-terminus of CLN2 of rat origin.

PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-34867 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

STORAGE

Store at 4° C, ****DO NOT FREEZE****. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

APPLICATIONS

CLN2 (L-12) is recommended for detection of CLN2 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500), immunohistochemistry (including paraffin-embedded sections) (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for CLN2 siRNA (h): sc-45578, CLN2 siRNA (m): sc-45579, CLN2 shRNA Plasmid (h): sc-45578-SH, CLN2 shRNA Plasmid (m): sc-45579-SH, CLN2 shRNA (h) Lentiviral Particles: sc-45578-V and CLN2 shRNA (m) Lentiviral Particles: sc-45579-V.

Molecular Weight of CLN2 precursor: 68 kDa.

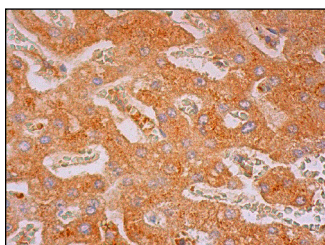
Molecular Weight of mature CLN2: 48 kDa.

Positive Controls: mouse heart extract: sc-2254.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941. 3) Immunohistochemistry: use ImmunoCruz™: sc-2053 or ABC: sc-2023 goat IgG Staining Systems.

DATA



CLN2 (L-12): sc-34867. Immunoperoxidase staining of formalin fixed, paraffin-embedded human liver tissue showing cytoplasmic staining of hepatocytes.

RESEARCH USE

For research use only, not for use in diagnostic procedures.



Try **CLN2 (G-3): sc-393961** or **CLN2 (D-11): sc-365838**, our highly recommended monoclonal alternatives to CLN2 (L-12).