SANTA CRUZ BIOTECHNOLOGY, INC.

CLN2 (D-11): sc-365838

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

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.

SOURCE

CLN2 (D-11) is a mouse monoclonal antibody raised against amino acids 264-563 mapping at the C-terminus of CLN2 of human origin.

PRODUCT

Each vial contains 200 µg IgG_1 kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

STORAGE

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

PROTOCOLS

See our web site at www.scbt.com for detailed protocols and support products.

APPLICATIONS

CLN2 (D-11) is recommended for detection of CLN2 of human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (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 shRNA Plasmid (h): sc-45578-SH and CLN2 shRNA (h) Lentiviral Particles: sc-45578-V.

Molecular Weight of CLN2 precursor: 68 kDa.

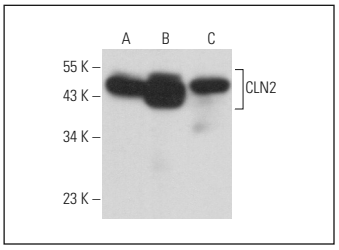
Molecular Weight of mature CLN2: 48 kDa.

Positive Controls: A-431 whole cell lysate: sc-2201, CCRF-CEM cell lysate: sc-2225 or Hep G2 cell lysate: sc-2227.

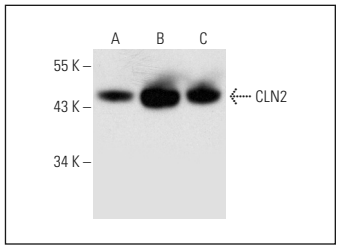
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

DATA



CLN2 (D-11): sc-365838. Western blot analysis of CLN2 expression in Hep G2 (**A**), THP-1 (**B**) and U-87 MG (**C**) whole cell lysates.



CLN2 (D-11): sc-365838. Western blot analysis of CLN2 expression in CCRF-CEM (**A**), Hep G2 (**B**) and A-431 (**C**) whole cell lysates.

SELECT PRODUCT CITATIONS

1. Ghosh, A., et al. 2012. Gemfibrozil and fenofibrate, food and drug administration-approved lipid-lowering drugs, up-regulate tripeptidyl-peptidase 1 in brain cells via peroxisome proliferator-activated receptor α: implications for late infantile Batten disease therapy. J. Biol. Chem. 287: 38922-38935.

RESEARCH USE

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