

# CLN1 (C-18): sc-21258

## BACKGROUND

CLN1 (ceroid lipofuscinosis 1), also known as PPT, INCL or PPT1 (palmitoyl-protein thioesterase 1), is a 306 amino acid glycosylated protein that localizes to lysosome and is a member of the palmitoyl-protein thioesterase family. CLN1 functions to remove thioester-linked fatty acyl groups from a variety of substrates, such as as palmitate, from modified cysteine residues in proteins or peptides during lysosomal degradation. Defects in the gene encoding CLN1 are a cause of infantile neuronal ceroid lipofuscinosis 1 (CLN1 or INCL) and neuronal ceroid lipofuscinosis 4 (CLN4). Neuronal ceroid lipofuscinoses are progressive neurodegenerative, lysosomal storage diseases characterized by intracellular accumulation of autofluorescent liposomal material, with clinical symptoms including seizures, dementia, visual loss and/or cerebral atrophy.

## REFERENCES

1. Vesa, J., et al. 1995. Mutations in the palmitoyl protein thioesterase gene causing infantile neuronal ceroid lipofuscinosis. *Nature* 376: 584-587.
2. Schriener, J.E., et al. 1996. cDNA and genomic cloning of human palmitoyl-protein thioesterase (PPT), the enzyme defective in infantile neuronal ceroid lipofuscinosis. *Genomics* 34: 317-322.
3. Crews, C.M., et al. 1996. Didemnin binds to the protein palmitoyl thioesterase responsible for infantile neuronal ceroid lipofuscinosis. *Proc. Natl. Acad. Sci. USA* 93: 4316-4319.
4. van Diggelen, O.P., et al. 2001. Adult neuronal ceroid lipofuscinosis with palmitoyl-protein thioesterase deficiency: first adult-onset patients of a childhood disease. *Ann. Neurol.* 50: 269-272.
5. Gupta, P., et al. 2001. Disruption of PPT1 or PPT2 causes neuronal ceroid lipofuscinosis in knockout mice. *Proc. Natl. Acad. Sci. USA* 98: 13566-13571.
6. Taschner, P.E., et al. 2005. From gene to disease; from CLN1, CLN2 and CLN3 to neuronal ceroid lipofuscinosis. *Ned. Tijdschr. Geneesk.* 149: 300-303.
7. Tsukamoto, T., et al. 2006. Overexpression in colorectal carcinoma of two lysosomal enzymes, CLN2 and CLN1, involved in neuronal ceroid lipofuscinosis. *Cancer* 106: 1489-1497.

## CHROMOSOMAL LOCATION

Genetic locus: PPT1 (human) mapping to 1p34.2; Ppt1 (mouse) mapping to 4 D2.2.

## STORAGE

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

## SOURCE

CLN1 (C-18) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the C-terminus of CLN1 of human 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-21258 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

## APPLICATIONS

CLN1 (C-18) is recommended for detection of CLN1 of human and, to a lesser extent, mouse and rat 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).

CLN1 (C-18) is also recommended for detection of CLN1 in additional species, including equine, canine and bovine.

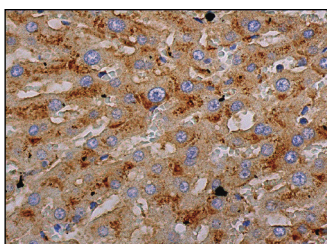
Suitable for use as control antibody for CLN1 siRNA (h): sc-105216, CLN1 siRNA (m): sc-142398, CLN1 shRNA Plasmid (h): sc-105216-SH, CLN1 shRNA Plasmid (m): sc-142398-SH, CLN1 shRNA (h) Lentiviral Particles: sc-105216-V and CLN1 shRNA (m) Lentiviral Particles: sc-142398-V.

Molecular Weight of glycosylated PPT1 doublet: 37 kDa.

## 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



CLN1 (C-18): sc-21258. 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.