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SANTA CRUZ BIOTECHNOLOGY, INC.

CLN1 siRNA (m): sc-142398

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. Schriner, 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. Geneeskd. 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 (mouse) mapping to 4 D2.2.

PRODUCT

CLN1 siRNA (m) is a pool of 3 target-specific 19-25 nt siRNAs designed to knock down gene expression. Each vial contains 3.3 nmol of lyophilized siRNA, sufficient for a 10 µM solution once resuspended using protocol below. Suitable for 50-100 transfections. Also see CLN1 shRNA Plasmid (m): sc-142398-SH and CLN1 shRNA (m) Lentiviral Particles: sc-142398-V as alternate gene silencing products.

For independent verification of CLN1 (m) gene silencing results, we also provide the individual siRNA duplex components. Each is available as 3.3 nmol of lyophilized siRNA. These include: sc-142398A, sc-142398B and sc-142398C.

STORAGE AND RESUSPENSION

Store lyophilized siRNA duplex at -20° C with desiccant. Stable for at least one year from the date of shipment. Once resuspended, store at -20° C, avoid contact with RNAses and repeated freeze thaw cycles.

Resuspend lyophilized siRNA duplex in 330 µl of the RNAse-free water provided. Resuspension of the siRNA duplex in 330 µl of RNAse-free water makes a 10 µM solution in a 10 µM Tris-HCl, pH 8.0, 20 mM NaCl, 1 mM EDTA buffered solution.

APPLICATIONS

CLN1 siRNA (m) is recommended for the inhibition of CLN1 expression in mouse cells.

SUPPORT REAGENTS

For optimal siRNA transfection efficiency, Santa Cruz Biotechnology's siRNA Transfection Reagent: sc-29528 (0.3 ml), siRNA Transfection Medium: sc-36868 (20 ml) and siRNA Dilution Buffer: sc-29527 (1.5 ml) are recommended. Control siRNAs or Fluorescein Conjugated Control siRNAs are available as 10 µM in 66 µl. Each contain a scrambled sequence that will not lead to the specific degradation of any known cellular mRNA. Fluorescein Conjugated Control siRNAs include: sc-36869, sc-44239, sc-44240 and sc-44241. Control siRNAs include: sc-37007, sc-44230, sc-44231, sc-44232, sc-44233, sc-44234, sc-44235, sc-44236, sc-44237 and sc-44238.

RT-PCR REAGENTS

Semi-quantitative RT-PCR may be performed to monitor CLN1 gene expression knockdown using RT-PCR Primer: CLN1 (m)-PR: sc-142398-PR (20 µl). Annealing temperature for the primers should be 55-60° C and the extension temperature should be 68-72° C.

SELECT PRODUCT CITATIONS

1. Sharma, G., et al. 2020. PPT1 inhibition enhances the anti-tumor activity of anti-PD-1 antibody in melanoma. JCI Insight. E-published.

RESEARCH USE

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

PROTOCOLS

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