# SANTA CRUZ BIOTECHNOLOGY, INC.

# CLN3 siRNA (h): sc-60406



# BACKGROUND

CLN3 is a highly glycosylated, hydrophobic, 438-amino acid protein with six transmembrane domains. The CLN3 protein localizes to the lysosomal membrane and plays a role in lysosomal function. It may act as a chaperone involved in the folding and unfolding of other proteins, namely subunit C of the ATP synthase complex. Mutations in the CLN3 gene cause Batten disease, a recessively inherited neurodegenerative disorder of childhood caused by lysosomal accumulation of hydrophobic material, mainly ATP synthase subunit C. Batten disease is the most common form of a group of disorders known as neuronal ceroid lipofuscinoses (NCLs). Symptoms of Batten disease include progressive loss of vision, seizures, and psychomotor disturbances.

# REFERENCES

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- Fossale, E., et al. 2004. Membrane trafficking and mitochondrial abnormalities precede subunit C deposition in a cerebellar cell model of juvenile neuronal ceroid lipofuscinosis. BMC Neurosci. 5: 57.
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- Mole, S.E., et al. 2005. Correlations between genotype, ultrastructural morphology and clinical phenotype in the neuronal ceroid lipofuscinoses. Neurogenetics 6: 107-126.
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- Persaud-Sawin, D.A., et al. 2005. Cell death pathways in juvenile Batten disease. Apoptosis 10: 973-985.
- Kwon, J.M., et al. 2005. Novel CLN3 mutation predicted to cause complete loss of protein function does not modify the classical JNCL phenotype. Neurosci. Lett. 387: 111-114.
- Pontikis, C.C., et al. 2005. Thalamocortical neuron loss and localized astrocytosis in the CLN3<sup>Δex7/8</sup> knock-in mouse model of Batten disease. Neurobiol. Dis. 20: 823-836.

#### CHROMOSOMAL LOCATION

Genetic locus: CLN3 (human) mapping to 16p11.2.

### PRODUCT

CLN3 siRNA (h) 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  $\mu$ M solution once resuspended using protocol below. Suitable for 50-100 transfections. Also see CLN3 shRNA Plasmid (h): sc-60406-SH and CLN3 shRNA (h) Lentiviral Particles: sc-60406-V as alternate gene silencing products.

For independent verification of CLN3 (h) gene silencing results, we also provide the individual siRNA duplex components. Each is available as 3.3 nmol of lyophilized siRNA. These include: sc-60406A, sc-60406B and sc-60406C.

#### 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  $\mu$ l of the RNAse-free water provided. Resuspension of the siRNA duplex in 330  $\mu$ l of RNAse-free water makes a 10  $\mu$ M solution in a 10  $\mu$ M Tris-HCl, pH 8.0, 20 mM NaCl, 1 mM EDTA buffered solution.

## **APPLICATIONS**

 $\ensuremath{\mathsf{CLN3}}$  siRNA (h) is recommended for the inhibition of  $\ensuremath{\mathsf{CLN3}}$  expression in human 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  $\mu$ M in 66  $\mu$ l. 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.

### **GENE EXPRESSION MONITORING**

CLN3 (C-1): sc-398192 is recommended as a control antibody for monitoring of CLN3 gene expression knockdown by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000) or immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500).

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgG $\kappa$  BP-HRP: sc-516102 or m-IgG $\kappa$  BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker<sup>TM</sup> Molecular Weight Standards: sc-2035, UltraCruz<sup>®</sup> Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunofluorescence: use m-IgG $\kappa$  BP-FITC: sc-516140 or m-IgG $\kappa$  BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz<sup>®</sup> Mounting Medium: sc-24941 or UltraCruz<sup>®</sup> Hard-set Mounting Medium: sc-359850.

### **RT-PCR REAGENTS**

Semi-quantitative RT-PCR may be performed to monitor CLN3 gene expression knockdown using RT-PCR Primer: CLN3 (h)-PR: sc-60406-PR (20  $\mu$ l). Annealing temperature for the primers should be 55-60° C and the extension temperature should be 68-72° C.

#### **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.