SANTA CRUZ BIOTECHNOLOGY, INC.

CLN3 (H-140): sc-134452

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The Power to Question

BACKGROUND

CLN3 is a highly glycosylated, hydrophobic, 438-amino acid protein with 6 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

1. Online Mendelian Inheritance in Man, OMIM™. 2002. Johns Hopkins University, Baltimore, MD. MIM Number: 204200. World Wide Web URL: http://www.ncbi.nlm.nih.gov/omim/
2. 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.
3. Leman, A.R., et al. 2005. Gene symbol: CLN3. Disease: Juvenile neuronal ceroid lipofuscinosis (Batten disease). Hum. Genet. 116: 544.
4. Mole, S.E., et al. 2005. Correlations between genotype, ultrastructural morphology and clinical phenotype in the neuronal ceroid lipofuscinoses. Neurogenetics 6: 107-126.
5. Phillips, S.N., et al. 2005. CLN3, the protein associated with batten disease: structure, function and localization. J. Neurosci. Res. 79: 573-583.
6. Persaud-Sawin, D.A., et al. 2005. Cell death pathways in juvenile Batten disease. Apoptosis 10: 973-985.
7. 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.
8. Pontikis, C.C., et al. 2005. Thalamocortical neuron loss and localized astrocytosis in the Cln3Deltaex7/8 knock-in mouse model of Batten disease. Neurobiol. Dis. 20: 823-836.

CHROMOSOMAL LOCATION

Genetic locus: CLN3 (human) mapping to 16p11.2; Cln3 (mouse) mapping to 7 F3.

SOURCE

CLN3 (H-140) is a rabbit polyclonal antibody raised against amino acids 299-438 mapping at the C-terminus of CLN3 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.

APPLICATIONS

CLN3 (H-140) is recommended for detection of CLN3 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, 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).

CLN3 (H-140) is also recommended for detection of CLN3 in additional species, including equine, canine, bovine and porcine.

Suitable for use as control antibody for CLN3 siRNA (h): sc-60406, CLN3 siRNA (m): sc-60407, CLN3 shRNA Plasmid (h): sc-60406-SH, CLN3 shRNA Plasmid (m): sc-60407-SH, CLN3 shRNA (h) Lentiviral Particles: sc-60406-V and CLN3 shRNA (m) Lentiviral Particles: sc-60407-V.

Molecular Weight of CLN3: 50 kDa.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (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) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

STORAGE

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

RESEARCH USE

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

PROTOCOLS

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