

CLN5 (C-15): sc-49928

BACKGROUND

Neuronal ceroid-lipofuscinose (NCL), also designated Batten disease, comprises a group of recessively inherited, progressive neurodegenerative diseases found in children. NCL is characterized by atrophy of the brain and an accumulation of lysosome derived fluorescent bodies found in many cells, especially neurons. Symptoms of NCL include a failure of psychomotor development, seizures, impaired vision and premature death. The eight genes/proteins associated with NCL are designated CLN1-CLN8. Mutations in six of these genes results in a distinct type of NCL-disease; the six genes/proteins are CLN1 (encoding PPT1, a protein thiolesterase), CLN2 (encoding the serine protease TPP1), CLN3, CLN5, CLN6 and CLN8. A single base duplication mutation in dog and cow CLN5 has been shown to cause NCL.

REFERENCES

1. Nardocci, N. and Cardona, F. 1998. Neuronal ceroid lipofuscinoses: a review. *Ital. J. Neurol. Sci.* 19: 271-276.
2. Wisniewski, K.E., Kida, E., Connell, F. and Zhong, N. 2000. Neuronal ceroid lipofuscinoses: research update. *Neurol. Sci.* 21: S49-56.
3. Zhong, N. 2000. Neuronal ceroid lipofuscinoses and possible pathogenic mechanism. *Mol. Genet. Metab.* 71: 195-206.
4. Heinonen, O., Salonen, T., Jalanko, A., Peltonen, L. and Copp, A. 2000. CLN1 and CLN5, genes for infantile and variant late infantile neuronal ceroid lipofuscinoses, are expressed in the embryonic human brain. *J. Comp. Neurol.* 426: 406-412.
5. Wisniewski, K.E., Zhong, N. and Philippart, M. 2001. Pheno/genotypic correlations of neuronal ceroid lipofuscinoses. *Neurology* 57: 576-581.
6. Ranta, S., Savukoski, M., Santavuori, P. and Haltia, M. 2001. Studies of homogenous populations: CLN5 and CLN8. *Adv. Genet.* 45: 123-140.
7. Melville, S.A., Wilson, C.L., Chiang, C.S., Studdert, V.P., Lingaas, F. and Wilton, A.N. 2005. A mutation in canine CLN5 causes neuronal ceroid lipofuscinosis in Border Collie dogs. *Genomics* 86: 287-294.
8. Mole, S.E., Williams, R.E. and Goebel, H.H. 2005. Correlations between genotype, ultrastructural morphology and clinical phenotype in the neuronal ceroid lipofuscinoses. *Neurogenetics* 63: 107-126.
9. Pineda-Trujillo, N., Cornejo, W., Carrizosa, J., Wheeler, R.B., Munera, S., Valencia, A., Agudelo-Arango, J., Cogollo, A., Anderson, G., Bedoya, G., Mole, S.E. and Ruiz-Linares, A. 2005. A CLN5 mutation causing an atypical neuronal ceroid lipofuscinosis of juvenile onset. *Neurology* 64: 740-742.

CHROMOSOMAL LOCATION

Genetic locus: CLN5 (human) mapping to 13q22.3.

SOURCE

CLN5 (C-15) is an affinity purified goat polyclonal antibody raised against a peptide mapping at the C-terminus of CLN5 of human origin.

RESEARCH USE

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

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-49928 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

CLN5 (C-15) is recommended for detection of CLN5 of human 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) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for CLN5 siRNA (h): sc-60408, CLN5 shRNA Plasmid (h): sc-60408-SH and CLN5 shRNA (h) Lentiviral Particles: sc-60408-V.

Molecular Weight of CLN5: 60 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.

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 or our catalog for detailed protocols and support products.



Try **CLN5 (D-8): sc-374672**, our highly recommended monoclonal alternative to CLN5 (C-15).