SANTA CRUZ BIOTECHNOLOGY, INC.

CLN5 (T-14): sc-49931

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

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 (encodeing the serine protease TPP1), CLN3, CLN5, CLN6 and CLN8. A single base duplication mutation in canine and bovine 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-S56.
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.

CHROMOSOMAL LOCATION

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

SOURCE

CLN5 (T-14) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of CLN5 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-49931 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

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.

APPLICATIONS

CLN5 (T-14) 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).

CLN5 (T-14) is also recommended for detection of CLN5 in additional species, including equine, canine, bovine, porcine and avian.

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.

PROTOCOLS

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