SANTA CRUZ BIOTECHNOLOGY, INC.

CLN5 (H-168): sc-134808

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 (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., et al. 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., et al. 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., et al. 2001. Pheno/genotypic correlations of neuronal ceroid lipofuscinoses. Neurology 57: 576-581.
6. Ranta, S., et al. 2001. Studies of homogenous populations: CLN5 and CLN8. Adv. Genet. 45: 123-140.
7. Melville, S.A., et al. 2005. A mutation in canine CLN5 causes neuronal ceroid lipofuscinosis in Border collie dogs. Genomics 86: 287-294.
8. Mole, S.E., et al. 2005. Correlations between genotype, ultrastructural morphology and clinical phenotype in the neuronal ceroid lipofuscinoses. Neurogenetics 63: 107-126.

CHROMOSOMAL LOCATION

Genetic locus: CLN5 (human) mapping to 13q22.3, Cln5 (mouse) mapping to 14 E2.3.

SOURCE

CLN5 (H-168) is a rabbit polyclonal antibody raised against amino acids 45-212 mapping near the N-terminus 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.

STORAGE

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

APPLICATIONS

CLN5 (H-168) is recommended for detection of CLN5 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).

CLN5 (H-168) is also recommended for detection of CLN5 in additional species, including canine, bovine and porcine.

Suitable for use as control antibody for CLN5 siRNA (h): sc-60408, CLN5 siRNA (m): sc-142399, CLN5 shRNA Plasmid (h): sc-60408-SH, CLN5 shRNA Plasmid (m): sc-142399-SH, CLN5 shRNA (h) Lentiviral Particles: sc-60408-V and CLN5 shRNA (m) Lentiviral Particles: sc-142399-V.

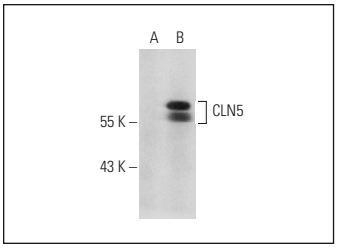
Molecular Weight of CLN5: 60 kDa.

Positive Controls: CLN5 (h5): 293T Lysate: sc-372537.

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.

DATA



CLN5 (H-168): sc-134808. Western blot analysis of CLN5 expression in non-transfected: sc-117752 (**A**) and human CLN5 transfected: sc-372537 (**B**) 293T whole cell lysates.

RESEARCH USE

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