SANTA CRUZ BIOTECHNOLOGY, INC.

CLN5 (D-8): sc-374672

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., 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.

CHROMOSOMAL LOCATION

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

SOURCE

CLN5 (D-8) is a mouse monoclonal antibody raised against amino acids 45-212 mapping near the N-terminus of CLN5 of human origin.

PRODUCT

Each vial contains 200 µg IgG_1 kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

CLN5 (D-8) is available conjugated to agarose (sc-374672 AC), 500 µg/ 0.25 ml agarose in 1 ml, for IP; to HRP (sc-374672 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-374672 PE), fluorescein (sc-374672 FITC), Alexa Fluor® 488 (sc-374672 AF488), Alexa Fluor® 546 (sc-374672 AF546), Alexa Fluor® 594 (sc-374672 AF594) or Alexa Fluor® 647 (sc-374672 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-374672 AF680) or Alexa Fluor® 790 (sc-374672 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

RESEARCH USE

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

APPLICATIONS

CLN5 (D-8) is recommended for detection of CLN5 of human origin by Western Blotting (starting dilution 1:100, 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), immunohistochemistry (including paraffin-embedded sections) (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.

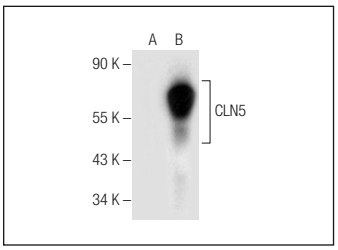
Molecular Weight of CLN5: 60 kDa.

Positive Controls: CLN5 (h4): 293T Lysate: sc-372321.

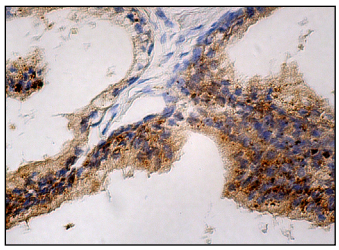
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 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 m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850. 4) Immunohistochemistry: use m-IgGκ BP-HRP: sc-516102 with DAB, 50X: sc-24982 and Immunohistomount: sc-45086, or Organo/Limonene Mount: sc-45087.

DATA



CLN5 (D-8): sc-374672. Western blot analysis of CLN5 expression in non-transfected: sc-117752 (**A**) and human CLN5 transfected: sc-372321 (**B**) 293T whole cell lysates.



CLN5 (D-8): sc-374672. Immunoperoxidase staining of formalin fixed, paraffin-embedded human prostate tissue showing cytoplasmic staining of glandular cells.

SELECT PRODUCT CITATIONS

1. Luo, S., et al. 2020. Functional analysis of a novel CLN5 mutation identified in a patient with neuronal ceroid lipofuscinosis. Front. Genet. 11: 536221.

STORAGE

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

Alexa Fluor® is a trademark of Molecular Probes, Inc., Oregon, USA