SANTA CRUZ BIOTECHNOLOGY, INC.

CLN8 (FL-286): sc-134453

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

CLN8, a 286 amino acid transmembrane protein, localizes mainly to the endoplasmic reticulum, but also partially to the ER-Golgi intermediate compartment (ERGIC). Mutations in the CLN8 gene cause neuronal ceroid lipofuscinosis 8 and progressive epilepsy with mental retardation (EPMR). Both disorders are forms of neuronal ceroid-lipofuscinose (NCL), a group of progressive neurodegenerative diseases found in children, characterized by failure of psychomotor development, impaired vision, seizures and premature death. The CLN8 protein is one of eight proteins in the CLN family, including CLN1-CLN7, which are associated with NCL.

REFERENCES

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2. Tyynela, J., et al. 2004. Hippocampal pathology in the human neuronal ceroid-lipofuscinoses: distinct patterns of storage deposition, neurodegeneration and glial activation. Brain Pathol. 14: 349-357.
3. Mole, S.E., et al. 2005. Correlations between genotype, ultrastructural morphology and clinical phenotype in the neuronal ceroid lipofuscinoses. Neurogenetics 6: 107-126.
4. Wendt, K.D., et al. 2005. Behavioral assessment in mouse models of neuronal ceroid lipofuscinosis using a light-cued T-maze. Behav. Brain Res. 161: 175-182.
5. Lonka, L., et al. 2005. The neuronal ceroid lipofuscinosis CLN8 gene expression is developmentally regulated in mouse brain and upregulated in the hippocampal kindling model of epilepsy. BMC Neurosci. 6: 27.
6. Katz, M.L., et al. 2005. A mutation in the CLN8 gene in English Setter dogs with neuronal ceroid-lipofuscinosis. Biochem. Biophys. Res. Commun. 327: 541-547.
7. Hermansson, M., et al. 2005. Mass spectrometric analysis reveals changes in phospholipid, neutral sphingolipid and sulfatide molecular species in progressive epilepsy with mental retardation, EPMR, brain: a case study. J. Neurochem. 95: 609-617.

CHROMOSOMAL LOCATION

Genetic locus: CLN8 (human) mapping to 8p23.3; Cln8 (mouse) mapping to 8 A1.1.

SOURCE

CLN8 (FL-286) is a rabbit polyclonal antibody raised against amino acids 1-286 representing full length CLN8 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.

RESEARCH USE

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

APPLICATIONS

CLN8 (FL-286) is recommended for detection of CLN8 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).

Suitable for use as control antibody for CLN8 siRNA (h): sc-60411, CLN8 siRNA (m): sc-60412, CLN8 shRNA Plasmid (h): sc-60411-SH, CLN8 shRNA Plasmid (m): sc-60412-SH, CLN8 shRNA (h) Lentiviral Particles: sc-60411-V and CLN8 shRNA (m) Lentiviral Particles: sc-60412-V.

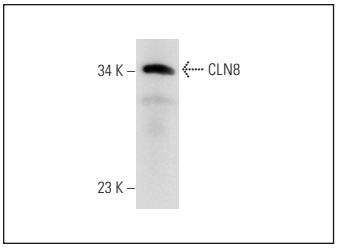
Molecular Weight of CLN8: 33 kDa.

Positive Controls: mouse brain extract: sc-2253.

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



CLN8 (FL-286): sc-134453. Western blot analysis of CLN8 expression in mouse brain tissue extract.

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.