SANTA CRUZ BIOTECHNOLOGY, INC.

CLN3 (E-19): sc-49626

BACKGROUND

CLN3 is a highly glycosylated, hydrophobic, 438 amino acid protein with six transmembrane domains. The CLN3 protein localizes to the lysosomal membrane and plays a role in lysosomal function. It may act as a chaperone involved in the folding and unfolding of other proteins, namely subunit C of the ATP synthase complex. Mutations in the CLN3 gene cause Batten disease, a recessively inherited neurodegenerative disorder of childhood caused by lysosomal accumulation of hydrophobic material, mainly ATP synthase subunit C. Batten disease is the most common form of a group of disorders known as neuronal ceroid lipofuscinoses (NCLs). Symptoms of Batten disease include progressive loss of vision, seizures and psychomotor disturbances.

REFERENCES

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4. Mole, S.E., et al. 2005. Correlations between genotype, ultrastructural morphology and clinical phenotype in the neuronal ceroid lipofuscinoses. Neurogenetics 6: 107-126.
5. Phillips, S.N., et al. 2005. CLN3, the protein associated with Batten disease: structure, function and localization. J. Neurosci. Res. 79: 573-583.
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CHROMOSOMAL LOCATION

Genetic locus: CLN3 (human) mapping to 16p12.1; Cln3 (mouse) mapping to 7 F3.

SOURCE

CLN3 (E-19) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of CLN3 of human origin.

STORAGE

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

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-49626 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

CLN3 (E-19) is recommended for detection of CLN3 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); only recommended for isoforms 2, 3 and 4.

Suitable for use as control antibody for CLN3 siRNA (h): sc-60406.

Molecular Weight of CLN3: 50 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.

RESEARCH USE

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

PROTOCOLS

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