

CLN3 (N-19): sc-49627

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

1. Online Mendelian Inheritance in Man, OMIM™. 2002. Johns Hopkins University, Baltimore, MD. MIM Number: 204200. World Wide Web URL: <http://www.ncbi.nlm.nih.gov/omim/>
2. Fossale, E., et al. 2004. Membrane trafficking and mitochondrial abnormalities precede subunit C deposition in a cerebellar cell model of juvenile neuronal ceroid lipofuscinosis. *BMC Neurosci.* 5: 57.
3. Leman, A.R., et al. 2005. Gene symbol: CLN3. Disease: Juvenile neuronal ceroid lipofuscinosis (Batten disease). *Hum. Genet.* 116: 544.
4. Mole, S.E., et al. 2005. Correlations between genotype, ultrastructural morphology and clinical phenotype in the neuronal ceroid lipofuscinoses. *Neurogenetics* 6: 107-126.
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CHROMOSOMAL LOCATION

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

SOURCE

CLN3 (N-19) is an affinity purified goat polyclonal antibody raised against a peptide mapping at the N-terminus of CLN3 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-49627 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

CLN3 (N-19) is recommended for detection of CLN3 of 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); recommended for all isoforms.

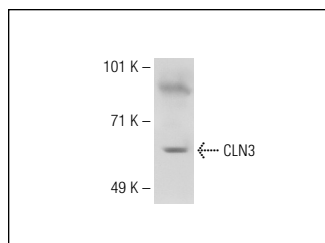
Suitable for use as control antibody for CLN3 siRNA (h): sc-60406, CLN3 shRNA Plasmid (h): sc-60406-SH and CLN3 shRNA (h) Lentiviral Particles: sc-60406-V.

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) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) 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.

DATA



CLN3 (N-19): sc-49627. Western blot analysis of CLN3 expression in HeLa whole cell lysate.

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.

MONOS
Satisfaction
Guaranteed

Try **CLN3 (C-1): sc-398192**, our highly recommended monoclonal alternative to CLN3 (N-19).