

CLN6 (E-19): sc-49631

BACKGROUND

CLN6, a 311 amino acid protein, has 7 predicted transmembrane domains and is conserved across vertebrates. The CLN6 protein localizes to the endoplasmic reticulum but contributes to lysosomal function. Mutations in the CLN6 gene cause variant late-onset infantile neuronal ceroid lipofuscinosis (vLINCL), a lysosomal storage disorder marked by progressive mental deterioration and blindness; part of a group of severe inherited neurodegenerative disorders affecting children wherein lysosomes accumulate storage material, causing the death of neurons. CLN6 is one of eight proteins, including CLN1-8, that are associated with NCL.

REFERENCES

1. Mole, S.E., et al. 2004. CLN6, which is associated with a lysosomal storage disease, is an endoplasmic reticulum protein. *Exp. Cell Res.* 298: 399-406.
2. Heine, C., et al. 2004. Defective endoplasmic reticulum-resident membrane protein CLN6 affects lysosomal degradation of endocytosed arylsulfatase A. *J. Biol. Chem.* 279: 22347-22352.
3. Oswald, M.J., et al. 2005. Glial activation spreads from specific cerebral foci and precedes neurodegeneration in presymptomatic ovine neuronal ceroid lipofuscinosis (CLN6). *Neurobiol. Dis.* 20: 49-63.
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. Siintola, E., et al. 2005. Two novel CLN6 mutations in variant late-infantile neuronal ceroid lipofuscinosis patients of Turkish origin. *Clin. Genet.* 68: 167-173.
6. Teixeira, C.A., et al. 2006. Gene expression profiling in vLINCL CLN6-deficient fibroblasts: Insights into pathobiology. *Biochim. Biophys. Acta* 1762: 637-646.

CHROMOSOMAL LOCATION

Genetic locus: CLN6 (human) mapping to 15q23; Cln6 (mouse) mapping to 9 B.

SOURCE

CLN6 (E-19) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of CLN6 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-49631 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

STORAGE

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

APPLICATIONS

CLN6 (E-19) is recommended for detection of CLN6 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); may cross-react with GRK5.

CLN6 (E-19) is also recommended for detection of CLN6 in additional species, including equine, canine, bovine, porcine and avian.

Suitable for use as control antibody for CLN6 siRNA (h): sc-60409, CLN6 siRNA (m): sc-60410, CLN6 shRNA Plasmid (h): sc-60409-SH, CLN6 shRNA Plasmid (m): sc-60410-SH, CLN6 shRNA (h) Lentiviral Particles: sc-60409-V and CLN6 shRNA (m) Lentiviral Particles: sc-60410-V.

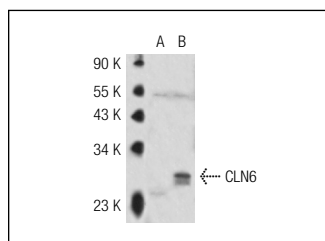
Molecular Weight of CLN6: 36 kDa.

Positive Controls: CLN6 (h2): 293T Lysate: sc-113803.

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



CLN6 (E-19): sc-49631. Western blot analysis of CLN6 expression in non-transfected: sc-117752 (A) and human CLN6 transfected: sc-113803 (B) 293T whole cell lysates.

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.