

CLN3 (C-1): sc-398192

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.

CHROMOSOMAL LOCATION

Genetic locus: CLN3 (human) mapping to 16p11.2; Cln3 (mouse) mapping to 7 F3.

SOURCE

CLN3 (C-1) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 13-40 at the N-terminus of CLN3 of human origin.

PRODUCT

Each vial contains 200 µg IgG_{2b} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

CLN3 (C-1) is available conjugated to agarose (sc-398192 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-398192 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-398192 PE), fluorescein (sc-398192 FITC), Alexa Fluor[®] 488 (sc-398192 AF488), Alexa Fluor[®] 546 (sc-398192 AF546), Alexa Fluor[®] 594 (sc-398192 AF594) or Alexa Fluor[®] 647 (sc-398192 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor[®] 680 (sc-398192 AF680) or Alexa Fluor[®] 790 (sc-398192 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

Blocking peptide available for competition studies, sc-398192 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

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APPLICATIONS

CLN3 (C-1) is recommended for detection of CLN3 of mouse, rat and 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) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for CLN3 siRNA (h): sc-60406, CLN3 siRNA (m): sc-60407, CLN3 shRNA Plasmid (h): sc-60406-SH, CLN3 shRNA Plasmid (m): sc-60407-SH, CLN3 shRNA (h) Lentiviral Particles: sc-60406-V and CLN3 shRNA (m) Lentiviral Particles: sc-60407-V.

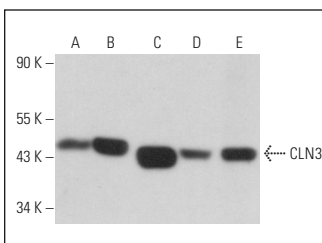
Molecular Weight of CLN3: 50 kDa.

Positive Controls: HeLa whole cell lysate: sc-2200, Jurkat whole cell lysate: sc-2204 or EOC 20 whole cell lysate: sc-364187.

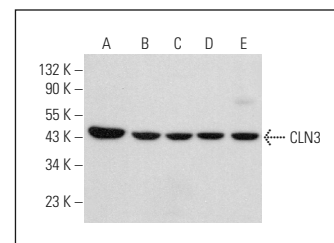
RECOMMENDED SUPPORT REAGENTS

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.

DATA



CLN3 (C-1): sc-398192. Western blot analysis of CLN3 expression in HeLa (A), Jurkat (B), Raji (C) and NCI-H929 (D) whole cell lysates and human brain tissue extract (E).



CLN3 (C-1): sc-398192. Western blot analysis of CLN3 expression in Jurkat (A), IMR-32 (B), Neuro-2A (C) and EOC 20 (D) whole cell lysates and rat brain tissue extract (E).

SELECT PRODUCT CITATIONS

1. Tang, C., et al. 2021. A human model of Batten disease shows role of CLN3 in phagocytosis at the photoreceptor-RPE interface. *Commun. Biol.* 4: 161.

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.