

ALS2CL (E-8): sc-377278

BACKGROUND

Mutations in the ALS2 gene result in a number of juvenile recessive motor neuron diseases (MNDs), including juvenile primary lateral sclerosis (JPLS), a recessive form of amyotrophic lateral sclerosis (ALS2), infantile onset ascending hereditary spastic paralysis (IAHSP) and a form of complicated hereditary spastic paraplegia (cHSP). The ALS2 gene encodes the Alsin protein. Alsin acts as a guanine nucleotide exchange factor for Rab5, a modulator of the endocytic pathway. Alsin is a cytosolic protein, which is associated with small, punctate membrane structures. Therefore Alsin may mediate membrane transport events, potentially linking endocytic processes and Actin cytoskeleton remodeling. The ALS2 C-terminal like protein (ALS2CL) also modulates Rab5 activity.

REFERENCES

1. Yang, Y., et al. 2001. The gene encoding Alsin, a protein with three guanine-nucleotide exchange factor domains, is mutated in a form of recessive amyotrophic lateral sclerosis. *Nat. Genet.* 29: 160-165.
2. Topp, J.D., et al. 2004. Alsin is a Rab5 and Rac1 guanine nucleotide exchange factor. *J. Biol. Chem.* 279: 24612-24623.
3. Hadano, S., et al. 2004. ALS2CL, the novel protein highly homologous to the carboxy-terminal half of ALS2, binds to Rab5 and modulates endosome dynamics. *FEBS Lett.* 575: 64-70.
4. Devon, R.S., et al. 2005. Cross-species characterization of the ALS2 gene and analysis of its pattern of expression in development and adulthood. *Neurobiol. Dis.* 18: 243-257.
5. Panzeri, C., et al. 2006. The first ALS2 missense mutation associated with JPLS reveals new aspects of Alsin biological function. *Brain* 129: 1710-1719.

CHROMOSOMAL LOCATION

Genetic locus: ALS2CL (human) mapping to 3p21.31.

SOURCE

ALS2CL (E-8) is a mouse monoclonal antibody raised against amino acids 765-840 mapping near the C-terminus of ALS2CL 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.

ALS2CL (E-8) is available conjugated to agarose (sc-377278 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-377278 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-377278 PE), fluorescein (sc-377278 FITC), Alexa Fluor® 488 (sc-377278 AF488), Alexa Fluor® 546 (sc-377278 AF546), Alexa Fluor® 594 (sc-377278 AF594) or Alexa Fluor® 647 (sc-377278 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-377278 AF680) or Alexa Fluor® 790 (sc-377278 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

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APPLICATIONS

ALS2CL (E-8) is recommended for detection of ALS2CL of 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 ALS2CL siRNA (h): sc-60156, ALS2CL shRNA Plasmid (h): sc-60156-SH and ALS2CL shRNA (h) Lentiviral Particles: sc-60156-V.

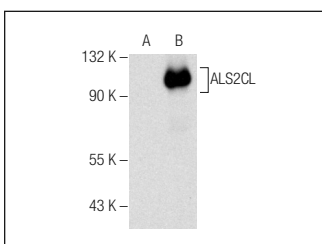
Molecular Weight of ALS2CL: 108 kDa.

Positive Controls: ALS2CL transfected 293T whole cell lysates.

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



ALS2CL (E-8): sc-377278. Western blot analysis of ALS2CL expression in non-transfected 293T (A) and human ALS2CL transfected 293T (B) whole cell lysates.

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.

PROTOCOLS

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