

ALS2CL (H-76): sc-134561

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 Rab 5, 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 Rab 5 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 Rab 5 and Rac 1 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 Rab 5 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: 1710-1709.
6. Matsuoka, M. and Nishimoto, I. 2006. Anti-ALS activity of Alsin, the product of the ALS2 gene and activity-dependent neurotrophic factor. *Neurodegener. Dis.* 2: 135-138.
7. Devon, R.S., et al. 2006. ALS2-deficient mice exhibit disturbances in endosome trafficking associated with motor behavioral abnormalities. *Proc. Natl. Acad. Sci. USA* 103: 9595-9600.

CHROMOSOMAL LOCATION

Genetic locus: ALS2CL (human) mapping to 3p21.31; Als2cl (mouse) mapping to 9 F3.

SOURCE

ALS2CL (H-76) is a rabbit polyclonal antibody raised against amino acids 765-840 mapping near the C-terminus of ALS2CL 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.

RESEARCH USE

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

APPLICATIONS

ALS2CL (H-76) is recommended for detection of ALS2CL 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).

ALS2CL (H-76) is also recommended for detection of ALS2CL in additional species, including equine, canine, bovine and porcine.

Suitable for use as control antibody for ALS2CL siRNA (h): sc-60156, ALS2CL siRNA (m): sc-60157, ALS2CL shRNA Plasmid (h): sc-60156-SH, ALS2CL shRNA Plasmid (m): sc-60157-SH, ALS2CL shRNA (h) Lentiviral Particles: sc-60156-V and ALS2CL shRNA (m) Lentiviral Particles: sc-60157-V.

Molecular Weight of ALS2CL: 108 kDa.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (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 goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

STORAGE

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

PROTOCOLS

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