

ALS2CL (N-16): sc-48448

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., Hentati, A., Deng, H.X., Dabbagh, O., Sasaki, T., Hirano, M., Hung, W.Y., Ouahchi, K., Yan, J., Azim, A.C., Cole, N., Gascon, G., Yagmour, A., Ben-Hamida, M., Pericak-Vance, M., Hentati, F. and Siddique, T. 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., Gray, N.W., Gerard, R.D. and Horazdovsky, B.F. 2004. Alsin is a Rab5 and Rac1 guanine nucleotide exchange factor. *J. Biol. Chem.* 279: 24612-24623.
3. Hadano, S., Otomo, A., Suzuki-Utsunomiya, K., Kunita, R., Yanagisawa, Y., Showguchi-Miyata, J., Mizumura, H. and Ikeda, J.E. 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., Schwab, C., Topp, J.D., Orban, P.C., Yang, Y.Z., Pape, T.D., Helm, J.R., Davidson, T.L., Rogers, D.A., Gros-Louis, F., Rouleau, G., Horazdovsky, B.F., Leavitt, B.R. and Hayden, M.R. 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., De Palma, C., Martinuzzi, A., Daga, A., De Polo, G., Bresolin, N., Miller, C.C., Tudor, E.L., Clementi, E. and Bassi, M.T. 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; Als2cl (mouse) mapping to 9 F3.

SOURCE

ALS2CL (N-16) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region 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.

Blocking peptide available for competition studies, sc-48448 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

ALS2CL (N-16) is recommended for detection of ALS2CL of mouse and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500), immunohistochemistry (including paraffin-embedded sections) (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

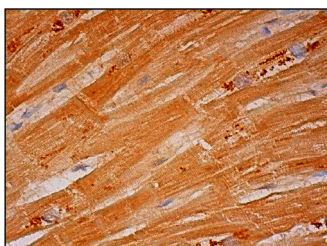
Suitable for use as control antibody for ALS2CL siRNA (h): sc-60156, ALS2CL siRNA (m): sc-60157, ALS2CL shRNA Plasmid (h): sc-60156-SH, ALS2CL shRNA Plasmid (m): sc-60157-SH, ALS2CL shRNA (h) Lentiviral Particles: sc-60156-V and ALS2CL shRNA (m) Lentiviral Particles: sc-60157-V.

Molecular Weight of ALS2CL: 108 kDa.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use 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) 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. 3) Immunohistochemistry: use ImmunoCruz™: sc-2053 or ABC: sc-2023 goat IgG Staining Systems.

DATA



ALS2CL (N-16): sc-48448. Immunoperoxidase staining of formalin fixed, paraffin-embedded human heart muscle tissue showing cytoplasmic staining of myocytes.

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 or our catalog for detailed protocols and support products.