



ALS2CR12 (M-15): sc-107396

BACKGROUND

Amyotrophic lateral sclerosis (ALS) is a fatal neurodegenerative disease characterized by progressive limb or bulbar weakness. Mutations in the ALS2 gene result in a number of juvenile recessive motor neuron diseases (MNDs), including juvenile primary lateral sclerosis (JPLS), infantile onset ascending hereditary spastic paralysis (IAHSP) and a form of complicated hereditary spastic paraplegia (cHSP). The ALS2 gene encodes the Alsin protein, which acts as a guanine nucleotide exchange factor for Rab 5, a modulator of the endocytic pathway. Alsin is a cytosolic protein that is associated with small, punctate membrane structures. Therefore Alsin may mediate membrane transport events, potentially linking endocytic processes and Actin cytoskeleton remodeling. ALS2CR12 (amyotrophic lateral sclerosis 2 (juvenile) chromosome region, candidate 12) is a 445 amino acid protein that may be involved in the regulation of GTPase 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: 1710-1719.
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. Jacquier, A., et al. 2006. Alsin/Rac 1 signaling controls survival and growth of spinal motoneurons. *Ann. Neurol.* 60: 105-117.
8. 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.
9. Cronin, S., et al. 2008. A genome-wide association study of sporadic ALS in a homogenous Irish population. *Hum. Mol. Genet.* 17: 768-774.

CHROMOSOMAL LOCATION

Genetic locus: Als2cr12 (mouse) mapping to 1 C1.3.

STORAGE

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

SOURCE

ALS2CR12 (M-15) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the C-terminus of ALS2CR12 of mouse 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-107396 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

ALS2CR12 (M-15) is recommended for detection of ALS2CR12 of mouse and rat 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) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000); non cross-reactive with other ALS2CR family members.

Suitable for use as control antibody for ALS2CR12 siRNA (m): sc-141032, ALS2CR12 shRNA Plasmid (m): sc-141032-SH and ALS2CR12 shRNA (m) Lentiviral Particles: sc-141032-V.

Molecular Weight of ALS2CR12: 52 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.

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.