SANTA CRUZ BIOTECHNOLOGY, INC.

ALS2CR4 (C-6): sc-514584

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

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, suggesting Alsin may mediate membrane transport events, potentially linking endocytic processes and actin cytoskeleton remodeling. ALS2CR4 (amyotrophic lateral sclerosis 2 (juvenile) chromosome region, candidate 4) is a 432 amino acid multi-pass membrane protein highly enriched in retina and localizes to photoreceptor outer segments, ciliary complex and postsynaptic outer plexiform layer. Encoded by a gene that maps to human chromosome 2q33.1, ALS2CR4 exists as two alternatively spliced isoforms.

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. 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.
4. Matsuoka, M., et al. 2005. Anti-ALS activity of alsin, the product of the ALS2 gene, and activity-dependent neurotrophic factor. Neurodegener. Dis. 2: 135-138.
5. Jacquier, A., et al. 2006. Alsin/Rac1 signaling controls survival and growth of spinal motoneurons. Ann. Neurol. 60: 105-117.

CHROMOSOMAL LOCATION

Genetic locus: TMEM237 (human) mapping to 2q33.1; Tmem237 (mouse) mapping to 1 C1.3.

SOURCE

ALS2CR4 (C-6) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 206-224 within an internal region of ALS2CR4 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.

Blocking peptide available for competition studies, sc-514584 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

APPLICATIONS

ALS2CR4 (C-6) is recommended for detection of ALS2CR4 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 ALS2CR4 siRNA (h): sc-95044, ALS2CR4 siRNA (m): sc-141034, ALS2CR4 shRNA Plasmid (h): sc-95044-SH, ALS2CR4 shRNA Plasmid (m): sc-141034-SH, ALS2CR4 shRNA (h) Lentiviral Particles: sc-95044-V and ALS2CR4 shRNA (m) Lentiviral Particles: sc-141034-V.

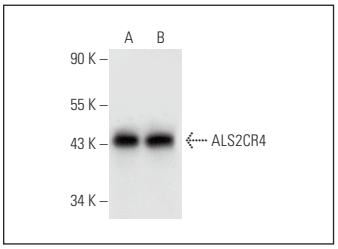
Molecular Weight of ALS2CR4: 48 kDa.

Positive Controls: human ovary extract: sc-363769 or human brain tissue extract.

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



ALS2CR4 (C-6): sc-514584. Western blot analysis of ALS2CR4 expression in human ovary (**A**) and human brain (**B**) tissue extracts.

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.