

ALS2CR4 (G-8): sc-514651

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.

CHROMOSOMAL LOCATION

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

SOURCE

ALS2CR4 (G-8) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 205-226 within an internal region of ALS2CR4 of human origin.

PRODUCT

Each vial contains 200 µg IgG₁ kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

ALS2CR4 (G-8) is available conjugated to agarose (sc-514651 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-514651 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-514651 PE), fluorescein (sc-514651 FITC), Alexa Fluor® 488 (sc-514651 AF488), Alexa Fluor® 546 (sc-514651 AF546), Alexa Fluor® 594 (sc-514651 AF594) or Alexa Fluor® 647 (sc-514651 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-514651 AF680) or Alexa Fluor® 790 (sc-514651 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

Blocking peptide available for competition studies, sc-514651 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

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APPLICATIONS

ALS2CR4 (G-8) 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.

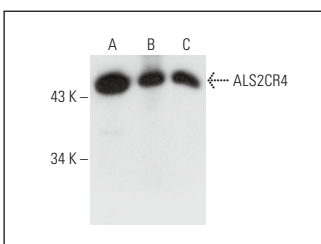
Molecular Weight of ALS2CR4: 48 kDa.

Positive Controls: HT-1080 whole cell lysate: sc-364183, human ovary extract: sc-363769 or human brain extract: sc-364375.

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 (G-8): sc-514651. Western blot analysis of ALS2CR4 expression in HT-1080 whole cell lysate (A) and human ovary (B) and human brain (C) 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.