

# ALS2CR7 (P-15): sc-103382

## BACKGROUND

Amotrophic 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 Rab5, 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. ALS2CR7 (amyotrophic lateral sclerosis 2 chromosomal region candidate gene 7 protein), also known as PFTK2, is a 384 amino acid protein belonging to the protein kinase superfamily. ALS2CR7 catalyzes the ATP-dependent phosphorylation of target proteins, thereby influencing signaling events throughout the cell. ALS2CR7 exists as three isoforms due to alternative splicing events.

## 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.

## CHROMOSOMAL LOCATION

Genetic locus: PFTK2 (human) mapping to 2q33.1; Pftk2 (mouse) mapping to 1 C1.3.

## SOURCE

ALS2CR7 (P-15) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the N-terminus of ALS2CR7 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-103382 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

## APPLICATIONS

ALS2CR7 (P-15) is recommended for detection of ALS2CR7 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); non cross-reactive with other ALS2CR family members.

ALS2CR7 (P-15) is also recommended for detection of ALS2CR7 in additional species, including equine, canine, bovine and porcine.

Suitable for use as control antibody for ALS2CR7 siRNA (h): sc-105057, ALS2CR7 siRNA (m): sc-105058, ALS2CR7 shRNA Plasmid (h): sc-105057-SH, ALS2CR7 shRNA Plasmid (m): sc-105058-SH, ALS2CR7 shRNA (h) Lentiviral Particles: sc-105057-V and ALS2CR7 shRNA (m) Lentiviral Particles: sc-105058-V.

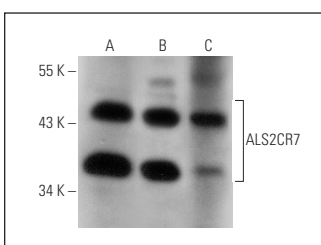
Molecular Weight of ALS2CR7: 44 kDa.

Positive Controls: HUV-EC-C whole cell lysate: sc-364180, human testis extract: sc-363781 or U-2 OS cell lysate: sc-2295.

## 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) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) 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.

## DATA



ALS2CR7 (P-15): sc-103382. Western blot analysis of ALS2CR7 expression in HUV-EC-C (A) and U-2 OS (B) whole cell lysates and human testis tissue extract (C).

## 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.