

# Alsin (2J10): sc-134256

## 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 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. The ALS2 C-terminal like protein (ALS2CL) also modulates Rab5 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/Rac1 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.

## CHROMOSOMAL LOCATION

Genetic locus: ALS2 (human) mapping to 2q33.1.

## SOURCE

Alsin (2J10) is a mouse monoclonal antibody raised against recombinant Alsin protein of human origin.

## PRODUCT

Each vial contains 100 µg IgG<sub>1</sub> kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

## RESEARCH USE

For research use only, not for use in diagnostic procedures.

## APPLICATIONS

Alsin (2J10) is recommended for detection of Alsin of 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)] and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for Alsin siRNA (h): sc-60158, Alsin shRNA Plasmid (h): sc-60158-SH and Alsin shRNA (h) Lentiviral Particles: sc-60158-V.

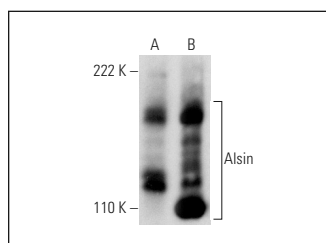
Molecular Weight of Alsin: 184 kDa.

Positive Controls: SH-SY5Y cell lysate: sc-3812, ACHN whole cell lysate: sc-364365 or human Alsin transfected 293T whole cell lysate.

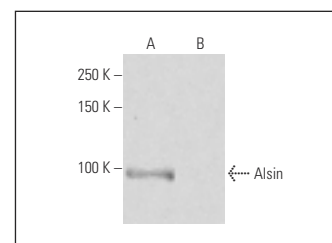
## RECOMMENDED SUPPORT REAGENTS

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).

## DATA



Alsin (2J10): sc-134256. Western blot analysis of Alsin expression in SH-SY5Y (A) and ACHN (B) whole cell lysates.



Alsin (2J10): sc-134256. Western blot analysis of Alsin expression in human Alsin transfected (A) and non-transfected (B) 293T whole cell lysates.

## STORAGE

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

## PROTOCOLS

See our web site at [www.scbt.com](http://www.scbt.com) for detailed protocols and support products.