

## Product Information

### Anti-ALS2CL

produced in rabbit, affinity isolated antibody

Product Number **SAB4200164**

### Product Description

Anti-ALS2CL is produced in rabbit using as the immunogen a synthetic peptide corresponding to a fragment of human ALS2CL (GeneID: 259173), conjugated to KLH. The corresponding sequence is identical in rat and mouse ALS2CL. The antibody is affinity-purified using the immunizing peptide immobilized on agarose.

Anti-ALS2CL specifically recognizes human ALS2CL. The antibody may be used in several immunochemical techniques including immunoblotting (~102 kDa). Detection of the ALS2CL band by immunoblotting is specifically inhibited by the ALS2CL immunizing peptide.

Amyotrophic lateral sclerosis (ALS) and primary lateral sclerosis (PLS) are heterogeneous neurological disorders that affect large motor neurons of the central nervous system (CNS). Loss-of function mutations in the ALS2 gene (also known as alsin) account for a number of recessive motor neuron diseases, including forms of ALS (ALS2), juvenile PLS (PLSJ), and hereditary spastic paraplegia (HSP).<sup>1-3</sup> ALS2 has Rab5-GEF activity and been shown to mediate the activation of Rab5 and Rac1/PACK1.<sup>3</sup>

ALS2CL (ALS2 C-terminal like) is an ALS2 homologous gene, encoding a 108 kDa protein. ALS2CL is present as a homo-dimeric form that can interact with ALS2 oligomer, resulting in a large ALS2-ALS2CL heteromeric complex. ALS2CL has been shown to interact and co-localize with ALS2 in membrane compartments.<sup>4</sup> In addition, ALS2CL suppresses the endosome enlargement induced by ALS2, resulting in an extensive perinuclear tubulo-membrane phenotype. In contrast to ALS2, ALS2CL exhibits a weak Rab5-GEF activity, but strongly binds to Rab5.<sup>5</sup> Co-expression of ALS2CL and Rab5 also results in a unique tubulation phenotype of endosome compartments and co-localization of ALS2CL and Rab5. It has been suggested that ALS2CL is an important factor modulating the ALS2 and Rab5-mediated endosome dynamics in the cell.

### Reagent

Supplied as a solution in 0.01 M phosphate buffered saline, pH 7.4, containing 15 mM sodium azide as a preservative.

Antibody concentration: ~1 mg/mL

### Precautions and Disclaimer

For R&D use only. Not for drug, household, or other uses. Please consult the Safety Data Sheet for information regarding hazards and safe handling practices.

### Storage/Stability

For continuous use, store at 2–8 °C for up to one month. For extended storage, freeze at –20 °C, in working aliquots. Repeated freezing and thawing, or storage in “frost-free” freezers, is not recommended. If slight turbidity occurs upon prolonged storage, clarify the solution by centrifugation before use. Working dilutions should be discarded if not used within 12 hours.

### Product Profile

**Immunoblotting:** a working antibody concentration of 0.5-1.0 µg/mL is recommended using HEK-293T cell lysates overexpressing human ALS2CL.

**Note:** In order to obtain best results in various techniques and preparations, it is recommended to determine optimal working dilutions by titration.

### References

1. Hadano, S., et al., *Nat. Genet.*, **29**, 166-173 (2001).
2. Yang, Y., et al., *Nat. Genet.*, **29**, 160-165 (2001).
3. Kunita, R., et al., *J. Biol. Chem.*, **282**, 16599-16611 (2007).
4. Suzuki-Utsunomiya, K., et al., *Biochem. Biophys. Res. Commun.*, **354**, 491-497 (2007).
5. Hadano, S., et al., *FEBS Lett.*, **575**, 64-70 (2004).

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