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Product Information

Anti-ALS2 (C-terminal)
produced in rabbit, affinity isolated antibody

Catalog Number **SAB4200350**

Product Description

Anti-ALS2 (C-terminal) is produced in rabbit using as immunogen a synthetic peptide corresponding to a sequence near the C-terminal region of rat ALS2 (GeneID: 363235), conjugated to KLH. The corresponding sequence is identical in mouse and highly conserved (single amino acid substitution) in human ALS2. The antibody is affinity-purified using the immunizing peptide immobilized on agarose.

Anti-ALS2 (C-terminal) specifically recognizes human, rat and mouse ALS2. The antibody may be used in various immunochemical techniques including immunoblotting (~185 kDa) and immunofluorescence. An additional weak band of ~150 kDa may be observed in some cell extracts. Detection of the ALS2 band by immunoblotting is specifically inhibited by the ALS2 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).[1-3] ALS2 contains several domains implicating roles in cell signaling and membrane dynamics. ALS2 consists of three putative guanine exchange factor (GEF) domains, the RCC1-like domain (RLD), the Dbl and pleckstrin homology (DH/PH) domain, and a vacuolar protein sorting 9 (VPS9) domain.[3] In addition, ALS2 contains eight consecutive membrane occupation and recognition nexus (MORN) motifs. ALS2 mRNA is ubiquitously expressed in the CNS and non-neuronal tissues, with the highest expression in the cerebellum and kidney. ALS2 has been shown to mediate the activation of Rab5 and Rac1/PACK1.[3,4] In the nervous system ALS2 is preferentially associated with the cytoplasmic side of the endosome membrane, it modulates endosome membrane trafficking and promotes neurite growth in neuronal cultures.

ALS2 knockout mice exhibit neurological deficits, altered endosome trafficking and degeneration of corticospinal neurons.[5,6]

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: ~1.5 mg/mL

Precautions and Disclaimer

This product is for R&D use only, not for drug, household, or other uses. Please consult the Material 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 in working aliquots at –20 °C. 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 1.5-3.0 µg/mL is recommended using HEK-293T cell lysates over-expressing human ALS2 or rat cerebellum extracts (S1 fraction).

Immunofluorescence: a working antibody concentration of 1-2 µg/mL is recommended using NIH3T3 cells.

Note: In order to obtain the best results using various techniques and preparations, we recommend determining the 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. Tudor, E.L., et al., *J. Biol. Chem.,* **280**, 34735-34740 (2005).
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6. Cai, H., et al., *Neurodegener. Dis.*, **5**, 359-366 (2008).

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