abcam

Product datasheet

Anti-Als2 antibody ab57550

1 Image

Overview

Product name	Anti-Als2 antibody
Description	Mouse monoclonal to Als2
Host species	Mouse
Tested applications	**Suitable for:** WB
Species reactivity	**Reacts with:** Human
Immunogen	Recombinant fragment: PSQDLKPVPE RCNQCSQLLI TMTDKEDHVI ISDSHCCPLG VTLTESQAEN HASTALSPST ETLDRQEEVF ENTLVANDQS VATELNAVSA QITSSDAMSS , corresponding to amino acids 221-321 of Human Als2 Run BLAST with ExPASy Run BLAST with NCBI

Properties

Form	Liquid
Storage instructions	Shipped at 4°C. Upon delivery aliquot and store at -20°C or -80°C. Avoid repeated freeze / thaw cycles.
Storage buffer	Preservative: None PBS, pH 7.2
Purity	Protein G purified
Clonality	Monoclonal
Isotype	IgG1
Light chain type	kappa

Applications

Our Abpromise guarantee covers the use of **ab57550** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Application	Abreviews	Notes

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WB		Use a concentration of 1 - 5 µg/ml. This antibody has only been tested in WB against the recombinant fragment used as immunogen. We have no data on the detection of endogenous protein.

Target

Function

May act as a GTPase regulator. Controls survival and growth of spinal motoneurons.

Involvement in disease

Defects in ALS2 are the cause of amyotrophic lateral sclerosis type 2 (ALS2) [MIM:205100]. ALS2 is a familial form of amyotrophic lateral sclerosis, a neurodegenerative disorder affecting upper and lower motor neurons and resulting in fatal paralysis. Sensory abnormalities are absent. Death usually occurs within 2 to 5 years. The etiology of amyotrophic lateral sclerosis is likely to be multifactorial, involving both genetic and environmental factors. The disease is inherited in 5-10% of cases leading to familial forms.
Defects in ALS2 are the cause of juvenile primary lateral sclerosis (JPLS) [MIM:606353]. JPLS is a neurodegenerative disorder which is closely related to but clinically distinct from amyotrophic lateral sclerosis. It is a progressive paralytic disorder which results from dysfunction of the upper motor neurons of the motor cortex while the lower neurons are unaffected.
Defects in ALS2 are the cause of infantile-onset ascending spastic paralysis (IAHSP) [MIM:607225]. IAHSP is characterized by progressive spasticity and weakness of limbs.

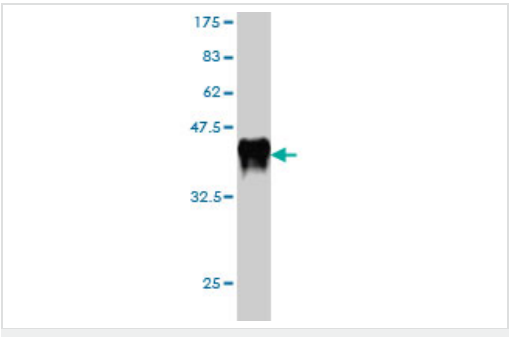
Sequence similarities

Contains 1 DH (DBL-homology) domain.
Contains 8 MORN repeats.
Contains 1 PH domain.
Contains 5 RCC1 repeats.
Contains 1 VPS9 domain.

Post-translational modifications

Phosphorylated upon DNA damage, probably by ATM or ATR.

Images



Western blot - Anti-Als2 antibody (ab57550)

Western blot against tagged recombinant protein immunogen using ab57550 Als2 antibody at 1ug/ml. Predicted band size of immunogen is 37 kDa

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