abcam

Product datasheet

Anti-Als2 antibody ab57550

1 Image

Overview

Product name	Anti-Als2 antibody
Description	Mouse monoclonal to Als2
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
WB		

Application notes	WB: Use at 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.

Not yet tested in other applications.
Optimal dilutions/concentrations should be determined by the end user.

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.

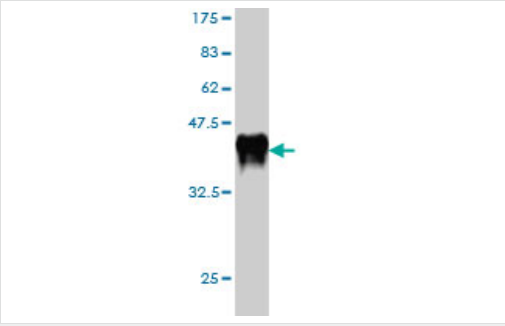
Sequence similarities

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 - 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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