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Rabbit Anti-ALS2 antibody

SL11709R

Product Name:	ALS2
Chinese Name:	肌萎缩侧索硬化蛋白2抗体
Alias:	ALS 2; ALS2; ALS2_HUMAN; ALS2CR6; Alsin; ALSJ; Amyotrophic lateral sclerosis 2 (juvenile); Amyotrophic lateral sclerosis 2 (juvenile) chromosome region candidate 6; Amyotrophic lateral sclerosis 2 chromosomal region candidate gene 6 protein; Amyotrophic lateral sclerosis 2 protein; Amyotrophic lateral sclerosis protein 2; FLJ31851; IAHSP; KIAA1563; MGC87187; PLSJ.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Pig,Cow,Horse,Rabbit,Sheep,
Applications:	ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	184kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human ALS2:1384-1440/1657
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
PubMed:	PubMed
Product Detail:	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 Rab5, 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 Rab 5 activity.

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

Subunit:
Forms a heteromeric complex with ALS2CL. Interacts with ALS2CL.

Post-translational modifications:
Phosphorylated upon DNA damage, probably by ATM or ATR.

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.

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

SWISS:
Q96Q42

Gene ID:
57679

	Database links: Entrez Gene: 57679 Human Entrez Gene: 363235 Rat GenBank: NM_020919 Human Omim: 606352 Human SwissProt: Q96Q42 Human SwissProt: P0C5Y8 Rat Unigene: 471096 Human Unigene: 621812 Human Unigene: 219733 Rat **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.