ARG22248
anti-Ataxin 1 antibody [S76-8]

Package: 50 µg
Store at: -20°C

Summary

Product Description	Mouse Monoclonal antibody [S76-8] recognizes Ataxin 1
Tested Reactivity	Hu, Ms, Rat
Tested Application	IHC-P, IP, WB
Specificity	Detects ~85kDa.
Host	Mouse
Clonality	Monoclonal
Clone	S76-8
Isotype	IgG2b
Target Name	Ataxin 1
Species	Mouse
Immunogen	Synthetic peptide around aa. 164-197 (ATTPSQRSQLEAYSTLLANMGSLSQAPGHKVEPP) of Mouse Ataxin 1. Rat: 100% identity (34/34 amino acids identical). Human: 88% identity (30/34 amino acids identical).
Conjugation	Un-conjugated
Alternate Names	SCA1; D6S504E; ATX1; Ataxin-1; Spinocerebellar ataxia type 1 protein

Application Instructions

Application table

Application	Dilution
IHC-P	Assay-dependent
IP	Assay-dependent
WB	1:1000

Application Note: * The dilutions indicate recommended starting dilutions and the optimal dilutions or concentrations should be determined by the scientist.

Properties

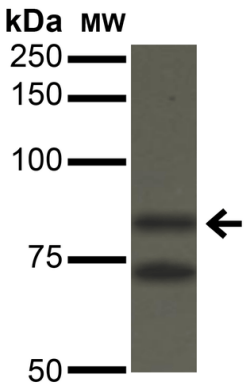
Form	Liquid
Purification	Purification with Protein G.
Buffer	PBS (pH 7.4), 0.1% Sodium azide and 50% Glycerol
Preservative	0.1% Sodium azide
Stabilizer	50% Glycerol
Concentration	1 mg/ml

Storage instruction	For continuous use, store undiluted antibody at 2-8°C for up to a week. For long-term storage, aliquot and store at -20°C. Storage in frost free freezers is not recommended. Avoid repeated freeze/thaw cycles. Suggest spin the vial prior to opening. The antibody solution should be gently mixed before use.
Note	For laboratory research only, not for drug, diagnostic or other use.

Bioinformation

Gene Symbol	Atxn1
Gene Full Name	ataxin 1
Background	The autosomal dominant cerebellar ataxias (ADCA) are a heterogeneous group of neurodegenerative disorders characterized by progressive degeneration of the cerebellum, brain stem and spinal cord. Clinically, ADCA has been divided into three groups: ADCA types I-III. ADCAI is genetically heterogeneous, with five genetic loci, designated spinocerebellar ataxia (SCA) 1, 2, 3, 4 and 6, being assigned to five different chromosomes. ADCAII, which always presents with retinal degeneration (SCA7), and ADCAIII often referred to as the `pure' cerebellar syndrome (SCA5), are most likely homogeneous disorders. Several SCA genes have been cloned and shown to contain CAG repeats in their coding regions. ADCA is caused by the expansion of the CAG repeats, producing an elongated polyglutamine tract in the corresponding protein. The expanded repeats are variable in size and unstable, usually increasing in size when transmitted to successive generations. The function of the ataxins is not known. This locus has been mapped to chromosome 6, and it has been determined that the diseased allele contains 41-81 CAG repeats, compared to 6-39 in the normal allele, and is associated with spinocerebellar ataxia type 1 (SCA1). At least two transcript variants encoding the same protein have been found for this gene. [provided by RefSeq, Jan 2010]
Function	Chromatin-binding factor that repress Notch signaling in the absence of Notch intracellular domain by acting as a CBF1 corepressor. Binds to the HEY promoter and might assist, along with NCOR2, RBPJ-mediated repression. Binds RNA in vitro. May be involved in RNA metabolism. [UniProt]
Calculated Mw	87 kDa
PTM	Ubiquitinated by UBE3A, leading to its degradation by the proteasome. The presence of expanded poly-Gln repeats in spinocerebellar ataxia 1 (SCA1) patients impairs ubiquitination and degradation, leading to accumulation of ATXN1 in neurons and subsequent toxicity. Phosphorylation at Ser-775 increases the pathogenicity of proteins with an expanded polyglutamine tract. Sumoylation is dependent on nuclear localization and phosphorylation at Ser-775. It is reduced in the presence of an expanded polyglutamine tract.
Cellular Localization	Cytoplasm, Nucleus

Images



ARG22248 anti-Ataxin 1 antibody [S76-8] WB image

Western blot: 15 µg of Monkey COS-1 cells transfected with Ataxin-1. Block: 2% BSA and 2% Skim Milk in 1X TBST. Primary Antibody: ARG22248 anti-Ataxin 1 antibody [S76-8] at 1:200 for 16 hours at 4°C. Secondary Antibody: Goat anti-Mouse IgG: HRP at 1:1000 for 1 hour RT. Color Development: ECL solution for 6 min in RT.