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ATXN1 Antibody

Purified Mouse Monoclonal Antibody
Catalog # AO1498a

Product Information

Application	WB, IHC, FC, ICC, E
Primary Accession	P54253
Reactivity	Human
Host	Mouse
Clonality	Monoclonal
Clone Names	2F5
Isotype	IgG1
Calculated MW	86923
Description	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. At least two transcript variants encoding the same protein have been found for this gene.Tissue specificity: Widely expressed throughout the body.
Immunogen	Purified recombinant fragment of human ATXN1 expressed in E. Coli.
Formulation	Ascitic fluid containing 0.03% sodium azide.

Additional Information

Gene ID	6310
Other Names	Ataxin-1, Spinocerebellar ataxia type 1 protein, ATXN1, ATX1, SCA1
Dilution	WB~~1/500 - 1/2000 IHC~~1/200 - 1/1000 FC~~1/200 - 1/400 ICC~~N/A E~~N/A
Storage	Maintain refrigerated at 2-8°C for up to 6 months. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.

Precautions	ATXN1 Antibody is for research use only and not for use in diagnostic or therapeutic procedures.

Protein Information

Name	ATXN1
Synonyms	ATX1, SCA1
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 (PubMed:21475249). In concert with CIC and ATXN1L, involved in brain development (By similarity).
Cellular Location	Cytoplasm. Nucleus Note=Colocalizes with USP7 in the nucleus
Tissue Location	Widely expressed throughout the body.

References

1. Nature. 2008 Apr 10;452(7188):713-8. 2. Biochem Biophys Res Commun. 2008 Jun 27;371(2):256-60. 3. Indian J Med Res. 2007 Nov;126(5):465-70.

Images

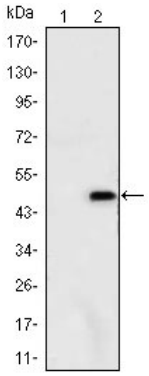


Figure 1: Western blot analysis using ATXN1 mAb against HEK293 (1) and ATXN1(AA: 645-815)-hIgGFc transfected HEK293 (2) cell lysate.

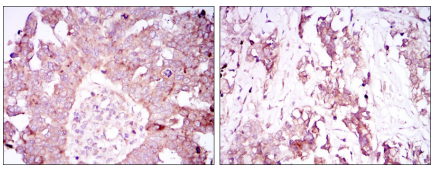
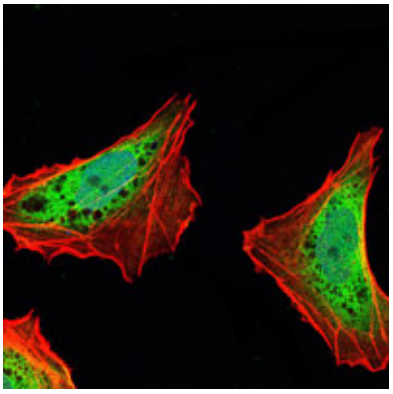


Figure 2: Immunohistochemical analysis of paraffin-embedded ovarian cancer tissues (left) and lung cancer tissues (right) using ATXN1 mouse mAb with DAB staining.

Figure 3: Immunofluorescence analysis of NTERA-2 cells using ATXN1 mouse mAb (green). Blue: DRAQ5 fluorescent DNA dye. Red: Actin filaments have been labeled with Alexa Fluor-555 phalloidin.



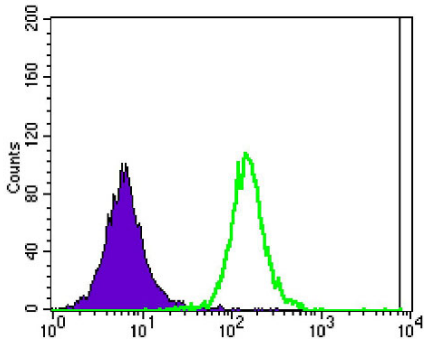


Figure 4: Flow cytometric analysis of Jurkat cells using ATXN1 mouse mAb (green) and negative control (purple).