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# ATXN1 Antibody

Purified Mouse Monoclonal Antibody
Catalog # AO1498a

## Product Information

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| **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

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| **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. |

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

## Protein Information

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| **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

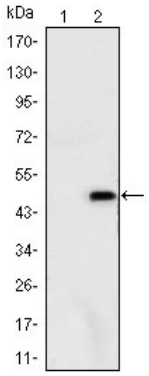



Figure 1: Western blot analysis using ATXN1 mAb against HEK293 (1) and ATXN1(AA: 645-815)-hIgGFc transfected HEK293 (2) cell lysate.

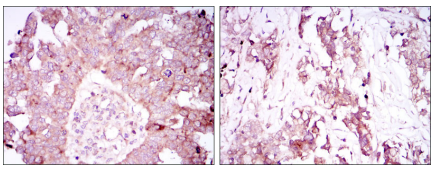
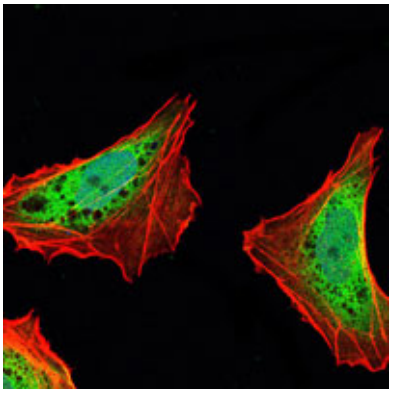


Figure 2: Immunohistochemical analysis of paraffin-embedded ovarian cancer tissues (left) and lung cancer tissues (right) using ATXN1 mouse mAb with DAB staining.

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.



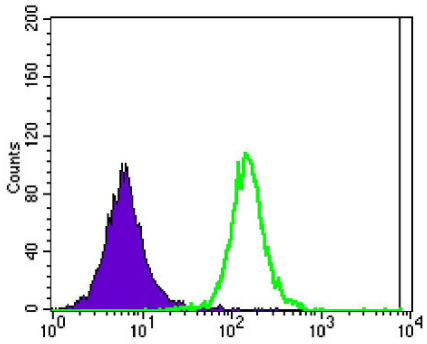



Figure 4: Flow cytometric analysis of Jurkat cells using ATXN1 mouse mAb (green) and negative control (purple).

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.