

Product name:	ATXN1 Mouse Monoclonal Antibody
Cat number:	MABN82209
Conjugate:	Unconjugated
Size:	100µL
Clone:	Monoclonal
Concentration:	1mg/ml
Host:	Mouse
Isotype:	Mouse IgG1
Immunogen:	Purified recombinant fragment of human ATXN1 (AA: 645-815) expressed in E. Coli.
Reactivity:	Human, Mouse
Applications:	WB 1:500-1:2000, ELISA 1:5000-1:20000, FC 1:200-1:400
Molecular Weight:	86.9kDa
Purification:	Affinity Purification
Form:	Liquid
Buffer:	Purified antibody in PBS with 0.05% sodium azide
Storage:	Store at 4°C short term. Aliquot and store at -20°C for 12 months. Avoid freeze/thaw cycles.
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 40-83 CAG repeats, compared to 6-39 in the normal allele, and is associated with spinocerebellar ataxia type 1 (SCA1). Alternative splicing results in multiple transcript variants, with one variant encoding multiple distinct proteins, ATXN1 and Alt-ATXN1, due to the use of overlapping alternate reading frames.