

ATXN1 Monoclonal Antibody

Description

Product type	Antibody
Code	BT-MCA4108
Host	Mouse
Isotype	Mouse IgG1
Size	100µL, 50µL
Immunogen	Purified recombinant fragment of human ATXN1 (AA: 645-815) expressed in E. Coli.
Mol wt	86.9kDa
Species reactivity	Human,Mouse
Clonality	Monoclonal
Recommended application	WB,FCM
Concentration	N/A
Full name	N/A
Synonyms	ATX1;SCA1;D6S504E

This product is for research use only, not for use in human, therapeutic or diagnostic procedure.

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. ADCA I is genetically heterogeneous, with five genetic loci, designated spinocerebellar ataxia (SCA) 1, 2, 3, 4 and 6, being assigned to five different chromosomes. ADCA II, which always presents with retinal degeneration (SCA7), and ADCA III 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.

Recommended Dilution

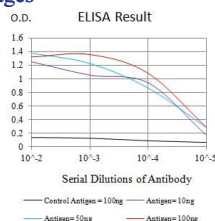
WB: 1:500 - 1:2000

FCM: 1:200 - 1:400

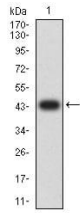
ELISA: 1:10000

Not yet tested in other applications.

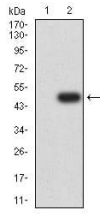
Images



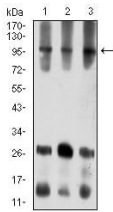
Black line: Control Antigen (100 ng); Purple line: Antigen (10ng); Blue line: Antigen (50 ng); Red line: Antigen (100 ng)



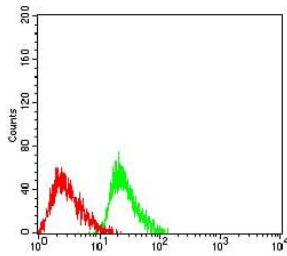
Western blot analysis using ATXN1 mAb against human ATXN1 recombinant protein.



Western blot analysis using ATXN1 mAb against HEK293 (1) and ATXN1-hlgGfc transfected HEK293 (2) cell lysate.



Western blot analysis using ATXN1 mouse mAb against COS7 (1), NIH/3T3 (2), and HL-60 (3) cell lysate.



Flow cytometric analysis of HL-60 cells using ATXN1 mouse mAb (green) and negative control (red).

Storage

Store at 4°C short term. Aliquot and store at -20°C long term.

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