

ATXN1 Rabbit pAb

Catalog No.: A6217

Basic Information

Observed MW

87kDa

Calculated MW

87kDa

Category

Primary antibody

Applications

ELISA, WB

Cross-Reactivity

Human, Mouse, Rat

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 Dilutions

WB 1:500 - 1:2000

Immunogen Information

Gene ID

6310

Swiss Prot

P54253

Immunogen

Recombinant fusion protein containing a sequence corresponding to amino acids 586-815 of human ATXN1 (NP_001121636.1).

Synonyms

ATX1; SCA1; D6S504E; ATXN1

Contact

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Product Information

Source

Rabbit

Isotype

IgG

Purification

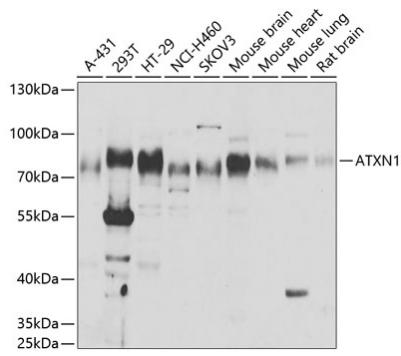
Affinity purification

Storage

Store at -20°C. Avoid freeze / thaw cycles.

Buffer: PBS with 0.02% sodium azide, 50% glycerol, pH 7.3.

Validation Data



Western blot analysis of extracts of various cell lines, using ATXN1 antibody (A6217) at 1:1000 dilution.
Secondary antibody: HRP Goat Anti-Rabbit IgG (H+L) (AS014) at 1:10000 dilution.
Lysates/proteins: 25µg per lane.
Blocking buffer: 3% nonfat dry milk in TBST.
Detection: ECL Basic Kit (RM00020).
Exposure time: 120s.