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ATXN1 Antibody (S776)

Affinity Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP2808A

Product Information

Application	IF, WB, E
Primary Accession	P54253
Reactivity	Human, Rat, Mouse
Host	Rabbit
Clonality	Polyclonal
Isotype	Rabbit IgG
Clone Names	RB16258
Calculated MW	86923
Antigen Region	754-781

Additional Information

Gene ID	6310
Other Names	Ataxin-1, Spinocerebellar ataxia type 1 protein, ATXN1, ATX1, SCA1
Target/Specificity	This ATXN1 antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide between 754-781 amino acids from human ATXN1.
Dilution	IF~~1:10~50 WB~~1:1000 E~~Use at an assay dependent concentration.
Format	Purified polyclonal antibody supplied in PBS with 0.09% (W/V) sodium azide. This antibody is purified through a protein A column, followed by peptide affinity purification.
Storage	Maintain refrigerated at 2-8°C for up to 2 weeks. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.
Precautions	ATXN1 Antibody (S776) 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.

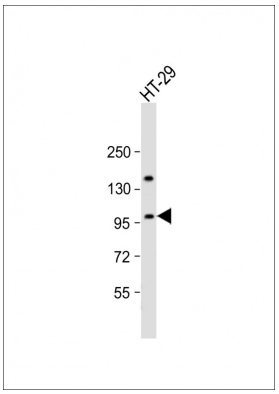
Background

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.

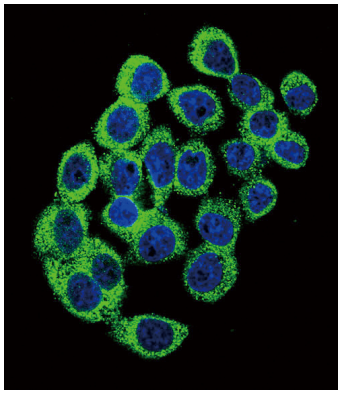
References

Hong,S., Biochem. Biophys. Res. Commun. 371 (2), 256-260 (2008)
Lim,J., Nature 452 (7188), 713-718 (2008)
Krol,H.A., PLoS ONE 3 (1), E1503 (2008)

Images



Anti-ATXN1 Antibody (S776) at 1:2000 dilution + HT-29 whole cell lysate Lysates/proteins at 20 µg per lane. Secondary Goat Anti-Rabbit IgG, (H+L), Peroxidase conjugated at 1/10000 dilution. Predicted band size :87kDa Blocking/Dilution buffer: 5% NFDM/TBST.



Confocal immunofluorescent analysis of ATXN1 Antibody (S776)(Cat#AP2808a) with hela cell followed by Alexa Fluor 488-conjugated goat anti-rabbit lgG (green). DAPI was used to stain the cell nuclear (blue).