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ATXN3 Antibody (Center)

Affinity Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP9244c

Product Information

Application	WB, IHC-P, FC, E
Primary Accession	P54252
Reactivity	Human
Predicted	Mouse, Rat, Bovine, Canine, Rabbit
Host	Rabbit
Clonality	Polyclonal
Isotype	Rabbit IgG
Clone Names	RB23951
Calculated MW	41250
Antigen Region	261-288

Additional Information

Gene ID	4287
Other Names	Ataxin-3, Machado-Joseph disease protein 1, Spinocerebellar ataxia type 3 protein, ATXN3, ATX3, MJD, MJD1, SCA3
Target/Specificity	This ATXN3 antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide between 261-288 amino acids from the Central region of human ATXN3.
Dilution	WB~~1:1000 IHC-P~~1:100~500 FC~~1:10~50 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	ATXN3 Antibody (Center) is for research use only and not for use in diagnostic or therapeutic procedures.

Protein Information

Name	ATXN3 {ECO:0000303|PubMed:33157014, ECO:0000312|HGNC:HGNC:7106}
Function	Deubiquitinating enzyme involved in protein homeostasis maintenance, transcription, cytoskeleton regulation, myogenesis and degradation of

misfolded chaperone substrates (PubMed:12297501, PubMed:16118278, PubMed:17696782, PubMed:23625928, PubMed:28445460, PubMed:33157014). Binds long polyubiquitin chains and trims them, while it has weak or no activity against chains of 4 or less ubiquitins (PubMed:17696782). Involved in degradation of misfolded chaperone substrates via its interaction with STUB1/CHIP: recruited to monoubiquitinated STUB1/CHIP, and restricts the length of ubiquitin chain attached to STUB1/CHIP substrates and preventing further chain extension (By similarity). Interacts with key regulators of transcription and represses transcription: acts as a histone-binding protein that regulates transcription (PubMed:12297501). Acts as a negative regulator of mTORC1 signaling in response to amino acid deprivation by mediating deubiquitination of RHEB, thereby promoting RHEB inactivation by the TSC-TBC complex (PubMed:33157014). Regulates autophagy via the deubiquitination of 'Lys-402' of BECN1 leading to the stabilization of BECN1 (PubMed:28445460).

Cellular Location

Nucleus matrix. Nucleus. Lysosome membrane; Peripheral membrane protein. Note=Predominantly nuclear, but not exclusively, inner nuclear matrix (PubMed:9580663). Recruited to lysosomal membrane in response to amino acid deprivation by the RagA/RRAGA-RagB/RRAGB complex (PubMed:33157014)

Tissue Location

Ubiquitous.

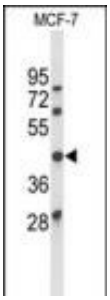
Background

ATXN3 was known as spinocerebellar ataxia-3, is an autosomal dominant neurologic disorder. The protein contains (CAG)n repeats in the coding region, and the expansion of these repeats from the normal 13-36 to 68-79 is one cause of Machado-Joseph disease. There is a negative correlation between the age of onset and CAG repeat numbers.

References

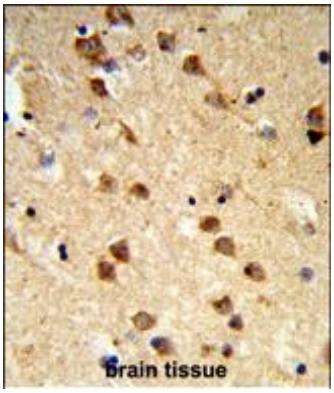
Reina,C.P., et.al, Hum. Mol. Genet. 19 (2), 235-249 (2010)
Jung,J., et.al, Hum. Mol. Genet. 18 (24), 4843-4852 (2009)

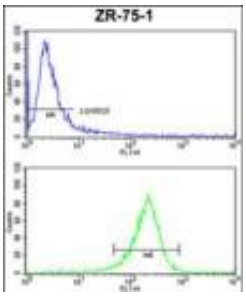
Images



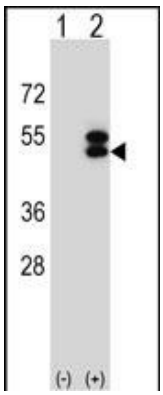
Western blot analysis of ATXN3 Antibody (Center) (Cat. #AP9244c) in MCF-7 cell line lysates (35ug/lane). ATXN3 (arrow) was detected using the purified Pab.

Formalin-fixed and paraffin-embedded human brain tissue reacted with ATXN3 Antibody (Center), which was peroxidase-conjugated to the secondary antibody, followed by DAB staining. This data demonstrates the use of this antibody for immunohistochemistry; clinical relevance has not been evaluated.





ATXN3 Antibody (Center) (Cat.#AP9244c) FC analysis of ZR-75-1 cells (bottom histogram) compared to a negative control cell (top histogram). FITC-conjugated goat-anti-rabbit secondary antibodies were used for the analysis.



Western blot analysis of ATXN3 (arrow) using rabbit polyclonal ATXN3 Antibody (Center) (Cat. #AP9244c). 293 cell lysates (2 ug/lane) either nontransfected (Lane 1) or transiently transfected (Lane 2) with the ATXN3 gene.

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