10320 Camino Santa Fe, Suite G
San Diego, CA 92121
Tel: 858.875.1900 Fax: 858.875.1999

Anti-Ataxin 3 Picoband Antibody

Catalog # ABO12113

Product Information

Application	WB, IHC-P
Primary Accession	P54252
Host	Rabbit
Reactivity	Human, Rat
Clonality	Polyclonal
Format	Lyophilized
Description	Rabbit IgG polyclonal antibody for Ataxin-3(ATXN3) detection. Tested with WB, IHC-P in Human;Rat.
Reconstitution	Add 0.2ml of distilled water will yield a concentration of 500ug/ml.

Additional Information

Gene ID	4287
Other Names	Ataxin-3, 3.4.19.12, Machado-Joseph disease protein 1, Spinocerebellar ataxia type 3 protein, ATXN3, ATX3, MJD, MJD1, SCA3
Calculated MW	41250
Application Details	Immunohistochemistry(Paraffin-embedded Section), 0.5-1 □g/ml, Human, By Heat Western blot, 0.1-0.5 □g/ml, Human, Rat
Subcellular Localization	Nucleus matrix . Predominantly nuclear, but not exclusively, inner nuclear matrix.
Tissue Specificity	Ubiquitous.
Source	Eukaryota
Protein Name	Ataxin-3
Contents	Each vial contains 5mg BSA, 0.9mg NaCl, 0.2mg Na2HPO4, 0.05mg NaN3.
Immunogen	A synthetic peptide corresponding to a sequence at the C-terminus of human Ataxin 3 (226-254aa EEDLQRALALSRQEIDMEDEEADLRRAIQ), different from the related mouse and rat sequences by two amino acids.
Purification	Immunogen affinity purified.
Cross Reactivity	No cross reactivity with other proteins

Storage	At -20˚C for one year. After r˚Constitution, at 4˚C for one month. It˚Can also be aliquotted and stored frozen at -20˚C for a longer time.Avoid repeated freezing and thawing.
Sequence Similarities	Contains 1 Josephin domain.

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.

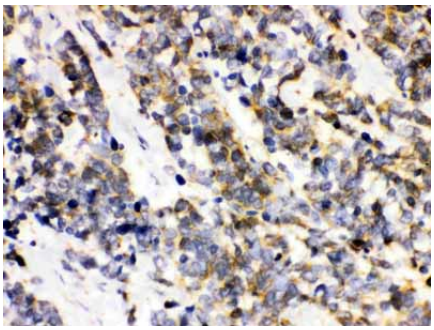
Background

ATXN3 (Ataxin 3), also known as AT3, MJD GENE, MJD1, SCA3 GENE, ATX3, JOS, Spinocerebellar ataxia-3, Machado-Joseph disease protein 1, is a protein that in humans is encoded by the ATXN3 gene. ATXN3 ranges in size from 360 to 374 amino acids. Using Northern blot analysis showed that ATXN3 mRNA was ubiquitously expressed in human tissues. They detected at least 4 ATXN3 transcripts of 1.4, 1.8, 4.5, and 7.5 kb and suggested that the different mRNA species probably result from differential splicing and polyadenylation. Machado-Joseph disease, also known as spinocerebellar ataxia-3, is an autosomal dominant neurologic disorder. The protein encoded by the ATXN3 gene contains (CAG)n repeats in the coding region, and the expansion of these repeats from the normal 13-36 to 68-79 is the cause of Machado-Joseph disease. There is an inverse correlation between the age of onset and CAG repeat numbers. Alternatively spliced transcript variants encoding different isoforms have been described for this gene. Ataxin-3 interacted with 2 human homologs of the yeast DNA repair protein RAD23, HHR23A (RAD23A) and HHR23B (RAD23B). Both normal and mutant ataxin-3 proteins interacted with the ubiquitin-like domain at the N terminus of the HHR23 proteins, which is a motif important for nucleotide excision repair. However, in HEK 293 cells, HHR23A was recruited to intranuclear inclusions formed by the mutant ataxin-3 through its interaction with ataxin-3.

Images



Anti- Ataxin 3 Picoband antibody, ABO12113, Western blottingAll lanes: Anti Ataxin 3 (ABO12113) at 0.5ug/mlLane 1: Rat Brain Tissue Lysate at 50ugLane 2: COLO320 Whole Cell Lysate at 40ugLane 3: HELA Whole Cell Lysate at 40ugPredicted bind size: 42KDObserved bind size: 42KD



Anti- Ataxin 3 Picoband antibody, ABO12113, IHC(P)IHC(P): Human Lung Cancer Tissue

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