

**Anti-Ataxin 3 Picoband Antibody**  
**Catalog # ABO12113****Specification**

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**Anti-Ataxin 3 Picoband Antibody - Product Information**

Application	WB, IHC-P
Primary Accession	<a href="#">P54252</a>
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.

**Anti-Ataxin 3 Picoband Antibody - 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**

41781 MW KDa

**Application Details**

Immunohistochemistry(Paraffin-embedded Section), 0.5-1 µg/ml, Human, By Heat<br>Western blot, 0.1-0.5 µg/ml, Human, Rat<br>

**Subcellular Localization**

Nucleus matrix . Predominantly nuclear, but not exclusively, inner nuclear matrix.

**Tissue Specificity**

Ubiquitous.

**Protein Name**

Ataxin-3

**Contents**

Each vial contains 5mg BSA, 0.9mg NaCl, 0.2mg Na<sub>2</sub>HPO<sub>4</sub>, 0.05mg NaN<sub>3</sub>.

**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 reconstitution, 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.

**Anti-Ataxin 3 Picoband Antibody - 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:<a href="http://www.uniprot.org/citations/12297501" target="\_blank">12297501</a>, PubMed:<a href="http://www.uniprot.org/citations/16118278" target="\_blank">16118278</a>, PubMed:<a href="http://www.uniprot.org/citations/17696782" target="\_blank">17696782</a>, PubMed:<a href="http://www.uniprot.org/citations/23625928" target="\_blank">23625928</a>, PubMed:<a href="http://www.uniprot.org/citations/28445460" target="\_blank">28445460</a>, PubMed:<a href="http://www.uniprot.org/citations/33157014" target="\_blank">33157014</a>). Binds long polyubiquitin chains and trims them, while it has weak or no activity against chains of 4 or less ubiquitins (PubMed:<a href="http://www.uniprot.org/citations/17696782" target="\_blank">17696782</a>). 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:<a href="http://www.uniprot.org/citations/12297501" target="\_blank">12297501</a>). 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:<a href="http://www.uniprot.org/citations/33157014" target="\_blank">33157014</a>). Regulates autophagy via the deubiquitination of 'Lys-402' of BECN1 leading to the stabilization of BECN1 (PubMed:<a href="http://www.uniprot.org/citations/28445460" target="\_blank">28445460</a>).

**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**

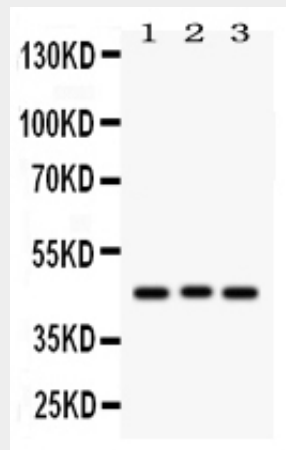
Ubiquitous.

**Anti-Ataxin 3 Picoband Antibody - Protocols**

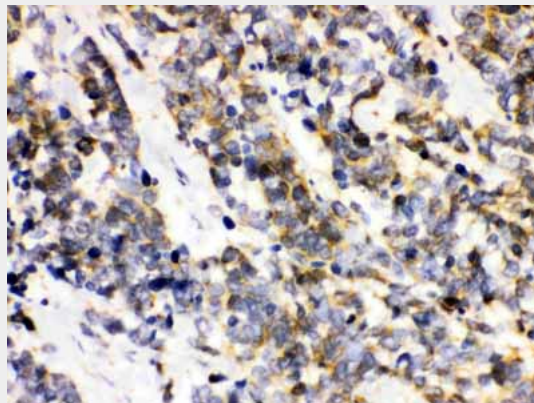
Provided below are standard protocols that you may find useful for product applications.

- [Western Blot](#)
- [Blocking Peptides](#)
- [Dot Blot](#)
- [Immunohistochemistry](#)
- [Immunofluorescence](#)
- [Immunoprecipitation](#)
- [Flow Cytometry](#)
- [Cell Culture](#)

#### Anti-Ataxin 3 Picoband Antibody - Images



Anti- Ataxin 3 Picoband antibody, ABO12113, Western blotting  
All lanes: Anti Ataxin 3 (ABO12113) at 0.5ug/ml  
Lane 1: Rat Brain Tissue Lysate at 50ug  
Lane 2: COLO320 Whole Cell Lysate at 40ug  
Lane 3: HELA Whole Cell Lysate at 40ug  
Predicted bind size: 42KD  
Observed bind size: 42KD



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

#### Anti-Ataxin 3 Picoband Antibody - 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)<sub>n</sub> 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.