

**Ataxin3 (MJD) Antibody (N-term) Blocking peptide**  
**Synthetic peptide**  
**Catalog # BP2181a****Specification**

---

**Ataxin3 (MJD) Antibody (N-term) Blocking peptide - Product Information**Primary Accession [P54252](#)**Ataxin3 (MJD) Antibody (N-term) Blocking peptide - 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**

The synthetic peptide sequence used to generate the antibody [AP2181a](/product/products/AP2181a) was selected from the N-term region of human MJD. A 10 to 100 fold molar excess to antibody is recommended. Precise conditions should be optimized for a particular assay.

**Format**

Peptides are lyophilized in a solid powder format. Peptides can be reconstituted in solution using the appropriate buffer as needed.

**Storage**

Maintain refrigerated at 2-8°C for up to 6 months. For long term storage store at -20°C.

**Precautions**

This product is for research use only. Not for use in diagnostic or therapeutic procedures.

**Ataxin3 (MJD) Antibody (N-term) Blocking peptide - 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](http://www.uniprot.org/citations/12297501), PubMed: [17696782](http://www.uniprot.org/citations/17696782), PubMed: [23625928](http://www.uniprot.org/citations/23625928), PubMed: [28445460](http://www.uniprot.org/citations/28445460), PubMed: [33157014](http://www.uniprot.org/citations/33157014), PubMed: [16118278](http://www.uniprot.org/citations/16118278)). Binds long polyubiquitin chains and trims them, while it has weak or no activity against chains of 4 or less ubiquitins (PubMed: [17696782](http://www.uniprot.org/citations/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:<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.

### Ataxin3 (MJD) Antibody (N-term) Blocking peptide - Protocols

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

- [Blocking Peptides](#)

### Ataxin3 (MJD) Antibody (N-term) Blocking peptide - Images

### Ataxin3 (MJD) Antibody (N-term) Blocking peptide - Background

Machado-Joseph disease is an autosomal dominant neurologic disorder, and is now known to be the same as previously described spinocerebellar ataxia-3. MJD protein (Ataxin-3) 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 a negative correlation between the age of onset and CAG repeat numbers. This protein interacts with key regulators (CBP, p300 and PCAF) of transcription and represses transcription, and also acts as a histone-binding protein that regulates transcription. MJD is a deubiquitinating enzyme.

### Ataxin3 (MJD) Antibody (N-term) Blocking peptide - References

Albrecht, M., et al., Proteins 50(2):355-370 (2003).Li, F., et al., J. Biol. Chem. 277(47):45004-45012 (2002).Strausberg, R.L., et al., Proc. Natl. Acad. Sci. U.S.A. 99(26):16899-16903 (2002).Ichikawa, Y., et al., J. Hum. Genet. 46(7):413-422 (2001).Tait, D., et al., Hum. Mol. Genet. 7(6):991-997 (1998).