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Ataxin3 (MJD) Antibody (C-term) Blocking peptide
Synthetic peptide
Catalog # BP2181b

Specification

Ataxin3 (MJD) Antibody (C-term) Blocking peptide - Product Information

Primary Accession P54252

Ataxin3 (MJD) Antibody (C-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 AP2181b was selected from the C-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 (C-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, 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.

Ataxin3 (MJD) Antibody (C-term) Blocking peptide - Protocols

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

- Blocking Peptides

Ataxin3 (MJD) Antibody (C-term) Blocking peptide - Images

Ataxin3 (MJD) Antibody (C-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)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 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 (C-term) Blocking peptide - References

Albrecht, M., et al., Eur. J. Biochem. 271(15):3155-3170 (2004).Michlewski, G., et al., J. Mol. Biol. 340(4):665-679 (2004).Li, Y., et al., Ann. Neurol. 56(1):124-129 (2004).Beuckmann, C.T., et al., J. Neurosci. 24(18):4469-4477 (2004).Berke, S.J., et al., J. Neurochem. 89(4):908-918 (2004).