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Ataxin 3 Rabbit pAb

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Catalog # AP56607

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	P54252
Reactivity	Rat, Human, Mouse
Predicted	Pig, Dog, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	41250
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human Ataxin 3
Epitope Specificity	51-150/364
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SUBCELLULAR LOCATION	Nucleus matrix. Predominantly nuclear, but not exclusively, inner nuclear matrix.
DISEASE	Defects in ATXN3 are the cause of spinocerebellar ataxia type 3 (SCA3) [MIM:109150]; also known as Machado-Joseph disease (MJD). Spinocerebellar ataxia is a clinically and genetically heterogeneous group of cerebellar disorders. Patients show progressive incoordination of gait and often poor coordination of hands, speech and eye movements, due to degeneration of the cerebellum with variable involvement of the brainstem and spinal cord. SCA3 belongs to the autosomal dominant cerebellar ataxias type I (ADCA I) which are characterized by cerebellar ataxia in combination with additional clinical features like optic atrophy, ophthalmoplegia, bulbar and extrapyramidal signs, peripheral neuropathy and dementia. The molecular defect in SCA3 is the a CAG repeat expansion in ATXN3 coding region. Longer expansions result in earlier onset and more severe clinical manifestations of the disease.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	Machado-Joseph disease, also known as spinocerebellar ataxia-3, is an autosomal dominant neurologic disorder. The protein encoded by this gene 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. Alternatively spliced transcript variants encoding different isoforms have been described for this gene. [provided by RefSeq, Sep 2009]

Additional Information

Gene ID	4287

Other Names	Ataxin-3, 3.4.19.12, Machado-Joseph disease protein 1, Spinocerebellar ataxia type 3 protein, ATXN3 {ECO:0000303|PubMed:33157014, ECO:0000312|HGNC:HGNC:7106}
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

Protein Information

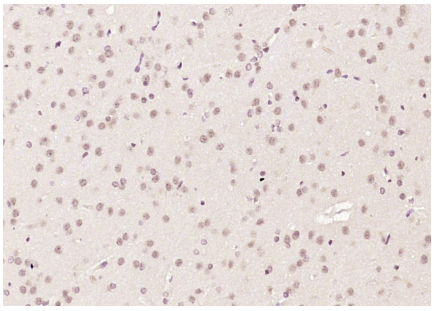
Name	ATXN3 {ECO:0000303|PubMed:33157014, ECO:0000312|HGNC:HGNC:7106}
Function	Deubiquitinating enzyme involved in protein homeostasis maintenance, transcription, cytoskeleton regulation, myogenesis and degradation of misfolded chaperone substrates (PubMed:12297501, PubMed:16118278, PubMed:17696782, PubMed:23625928, 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

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Images

Paraformaldehyde-fixed, paraffin embedded (rat brain); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (Ataxin 3) Polyclonal Antibody, Unconjugated (AP56607) at 1:200 overnight at 4°C, followed by operating according to SP Kit(Rabbit)



(sp-0023) instructionsand DAB staining.

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