

Ataxin 7 Rabbit pAb

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Catalog # AP54452

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	O15265
Reactivity	Rat, Mouse
Predicted	Pig, Dog, Human, Rabbit, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	95451
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human Ataxin 7
Epitope Specificity	301-400/892
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	Cytoplasmic (isoform b) and Nuclear (isoform a)
DISEASE	Defects in ATXN7 are the cause of spinocerebellar ataxia type 7 (SCA7) [MIM:164500]; also known as olivopontocerebellar atrophy III (OPCA III or OPCA3) or olivopontocerebellar atrophy with retinal degeneration. 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. SCA7 belongs to the autosomal dominant cerebellar ataxias type II (ADCA II) which are characterized by cerebellar ataxia with retinal degeneration and pigmentary macular dystrophy.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	The human ataxin-7 gene, also known as spinocerebellar ataxia 7 or SCA7, maps to chromosome 3p13-p12, has a 2,727-bp open reading frame, and encodes a 892 amino acid protein containing a nuclear localization signal and a polyglutamine tract (1,2). SCA7 is an autosomal dominant neurodegenerative disorder characterized by ataxia and selective neuronal cell loss caused by the expansion of a translated CAG repeat encoding a polyglutamine tract in ataxin-7, which is the SCA7 gene product (3,4). Ataxin-7 is expressed within neurons both affected and unaffected in SCA7 pathology with subcellular localization being variable depending upon the neuronal subtype (5). Polyglutamine expanded in ataxin-7 may carry out its pathogenic effects in the nucleus by altering the matrix-associated nuclear structure and/or by disrupting nucleolar function (6).

Additional Information

Gene ID	6314
Other Names	Ataxin-7, Spinocerebellar ataxia type 7 protein, ATXN7, SCA7 {ECO:0000303 PubMed:9288099}
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.

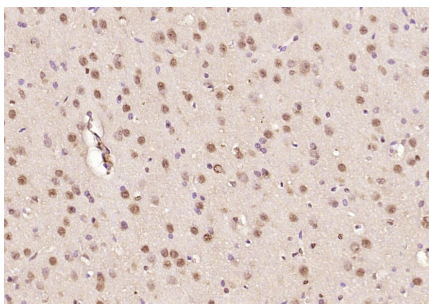
Protein Information

Name	ATXN7
Synonyms	SCA7 {ECO:0000303 PubMed:9288099}
Function	Acts as a component of the SAGA (aka STAGA) transcription coactivator-HAT complex (PubMed: 15932940 , PubMed: 18206972). Mediates the interaction of SAGA complex with the CRX and is involved in CRX- dependent gene activation (PubMed: 15932940 , PubMed: 18206972). Probably involved in tethering the deubiquitination module within the SAGA complex (PubMed: 24493646). Necessary for microtubule cytoskeleton stabilization (PubMed: 22100762). Involved in neurodegeneration (PubMed: 9288099).
Cellular Location	[Isoform a]: Nucleus. Nucleus, nucleolus. Nucleus matrix. Cytoplasm, cytoskeleton. Note=In addition to a diffuse distribution throughout the nucleus, it is associated with the nuclear matrix and the nucleolus (PubMed:10441328). It is able to shuttle between the nucleus and cytoplasm (PubMed:16314424)
Tissue Location	[Isoform a]: Isoform a is expressed in CNS, but is expressed predominantly in the peripheral tissues

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 7) Polyclonal Antibody, Unconjugated (AP54452) at 1:200 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructions and DAB staining.

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