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Rabbit Anti-ATXN3L antibody

SL4807R

Product Name:	ATXN3L
Chinese Name:	小脑脊髓共济失调蛋白3抗体
Alias:	ATX3L_HUMAN; ATXN3L; Machado-Joseph disease protein 1-like; MJDL; Putative ataxin-3-like protein.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,
Applications:	WB=1:500-2000IHC-P=1:400-800IHC-F=1:400-800IF=1:50-200（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	41kDa
Cellular localization:	The cell membrane
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human ATXN3L:251-355/355
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. 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.
PubMed:	PubMed
Product Detail:	Defects in ATXN3 are the cause of spinocerebellar ataxia type 3 (SCA3) ; 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.

Function:
Deubiquitinating enzyme that cleaves both 'Lys-48'-linked and 'Lys-63'-linked poly-ubiquitin chains (in vitro).

Subcellular Location:
Nucleus (By similarity).

Similarity:
Contains 1 Josephin domain.
Contains 2 UIM (ubiquitin-interacting motif) repeats.

SWISS:
Q9H3M9

Gene ID:
25814

Database links:

Entrez Gene: 25814Human

Entrez Gene: 170821Rat

Omim: 611150Human

SwissProt: Q9UBB4Human

SwissProt: Q9ER24Rat

Unigene: 475125Human

Unigene: 6524Rat

Important Note:
This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.