ATX3 (Phospho Ser256) Rabbit pAb

CatalogNo: YP1766

Key Features

Host Species
- Rabbit

Reactivity
- Human,Mouse,Rat

Applications
- WB

MW
- 40kD (Calculated)

Isotype
- IgG

Recommended Dilution Ratios

WB 1:500-2000

Storage

Storage*	-15°C to -25°C/1 year(Do not lower than -25°C)
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.

Basic Information

Clonality	Polyclonal

Immunogen Information

Immunogen	Synthesized peptide derived from human Ataxin-3 (Phospho-Ser256)
Specificity	This antibody detects endogenous levels of Ataxin-3 (Phospho-Ser256) at Human, Mouse,Rat.The name of modified sites may be influenced by many factors, such as species (the modified site was not originally found in human samples) and the change of protein sequence (the previous protein sequence is incomplete, and the protein sequence may be prolonged with the development of protein sequencing technology). When naming, we will use the "numbers" in historical reference to keep the sites consistent with the reports. The antibody binds to the following modification sequence (lowercase letters are modification sites):QLsMQ

Target Information

Gene name ATXN3 ATX3 MJD MJD1 SCA3

Protein Name Ataxin-3 (Phospho-Ser256)

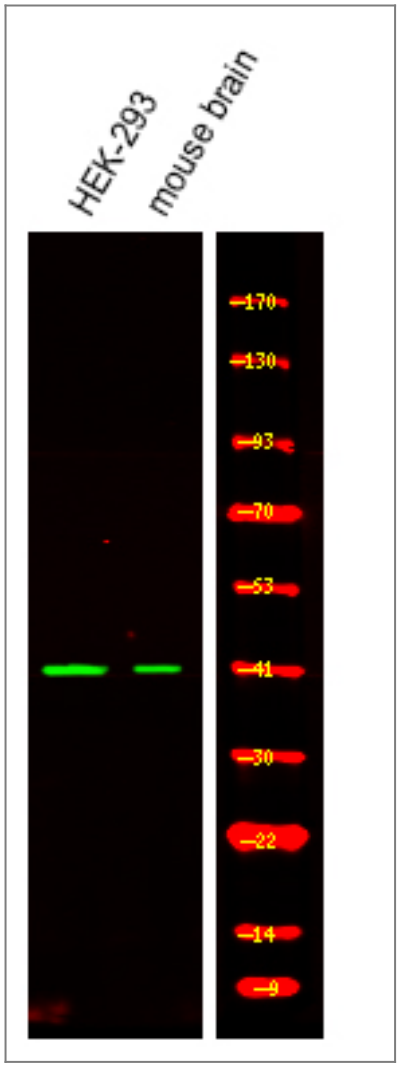
Organism	Gene ID	UniProt ID
Human	4287;	P54252;
Mouse	110616;	Q9CVD2;
Rat	60331;	O35815;

Cellular Localization Nucleus matrix . Nucleus . Predominantly nuclear, but not exclusively, inner nuclear matrix.

Tissue specificity Ubiquitous.

Function 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 ATX3 coding region. Longer expansions result in earlier onset and more severe clinical manifestations of the disease.,Function:Interacts with key regulators (CBP, p300 and PCAF) of transcription and represses transcription. Acts as a histone-binding protein that regulates transcription. Acts as a deubiquitinating enzyme.,polymorphism:The MJD1a allele carries a single nucleotide substitution in codon 349 generating a stop codon instead of a Tyr. In the Japanese population, the MJD1a allele seems to be significantly associated with Gln expansion.,polymorphism:The poly-Gln region of ATXN3 is highly polymorphic (14 to 41 repeats) in the normal population and is expanded to about 55-82 repeats in spinocerebellar ataxia 3 (SCA3) patients.,similarity:Contains 1 Josephin domain.,similarity:Contains 3 UIM (ubiquitin-interacting motif) repeats.,subcellular location:Predominantly nuclear, but not exclusively, inner nuclear matrix.,subunit:Interacts with DNA repair proteins RAD23A and RAD23B.,tissue specificity:Ubiquitous.,

Validation Data



Western Blot analysis of various,using primary antibody at 1:1000 dilution. Secondary antibody(catalog#:RS23920) was diluted at 1:10000

Contact information

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