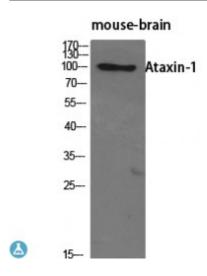


Anti-Ataxin-1 antibody



Description Rabbit polyclonal to Ataxin-1.

Model STJ91738

Host Rabbit

Reactivity Human, Mouse

Applications ELISA, IF, IHC

Immunogen Synthesized peptide derived from human Ataxin-1 around the non-

phosphorylation site of S776.

Immunogen Region 720-800 aa

Gene ID <u>6310</u>

Gene Symbol ATXN1

Dilution range IHC 1:100-1:300IF 1:200-1:1000ELISA 1:5000

Specificity Ataxin-1 Polyclonal Antibody detects endogenous levels of Ataxin-1 protein.

Tissue Specificity Widely expressed throughout the body.

Purification The antibody was affinity-purified from rabbit antiserum by affinity-

chromatography using epitope-specific immunogen.

Note For Research Use Only (RUO).

Protein Name Ataxin-1 Spinocerebellar ataxia type 1 protein

Molecular Weight 87 kDa

Clonality Polyclonal

Conjugation Unconjugated

Isotype IgG

Formulation Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.

Concentration 1 mg/ml

Storage Instruction Store at -20°C, and avoid repeat freeze-thaw cycles.

Database Links <u>HGNC:10548OMIM:164400</u>

Alternative Names Ataxin-1 Spinocerebellar ataxia type 1 protein

Function Chromatin-binding factor that repress Notch signaling in the absence of Notch

intracellular domain by acting as a CBF1 corepressor. Binds to the HEY promoter and might assist, along with NCOR2, RBPJ-mediated repression.

Binds RNA in vitro. May be involved in RNA metabolism.

Sequence and Domain Family The AXH domain is required for interaction with CIC.

Cellular Localization Cytoplasm Nucleus. Colocalizes with USP7 in the nucleus.

Post-translational Ubiquitinated by UBE3A, leading to its degradation by the proteasome. The **Modifications** presence of expanded poly-Gln repeats in spinocerebellar ataxia 1 (SCA1)

presence of expanded poly-Gln repeats in spinocerebellar ataxia 1 (SCA1) patients impairs ubiquitination and degradation, leading to accumulation of ATXN1 in neurons and subsequent toxicity. Phosphorylation at Ser-775 increases the pathogenicity of proteins with an expanded polyglutamine tract. Sumoylation is dependent on nuclear localization and phosphorylation at Ser-775. It is reduced in the presence of an expanded polyglutamine tract.

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