SANTA CRUZ BIOTECHNOLOGY, INC.

# Ataxin-3 (C-5): sc-393193

SANTA CRUZ BIOTECHNOLOGY

*The Power to Question*

## BACKGROUND

Autosomal dominant cerebellar ataxias are a group of neuro-degenerative disorders caused by unstable CAG repeat expansions encoding polyglutamine tracts. Proteins with long polyglutamine tracts have an increased tendency to aggregate, often forming ubiquitinated intranuclear inclusion bodies. Machado-Joseph disease (MJD)/spinocerebellar ataxia type 3 (SCA3) gene encodes Ataxin-3, which contains a polyglutamine stretch. Ataxin-3 is incorporated into most of the nuclear inclusions (NIs) and disappears from its normal cytoplasmic localization under pathological conditions in most neurons. However, in the early onset of SCA3, the association of a pathological form of Ataxin-3 with nuclear matrix alters Ataxin-3 conformation to expose the polyglutamine domain. In normal brain tissue, wild-type Ataxin-3 can also be localized within the ubiquitin-positive nuclear inclusion, the Marinesco body, under certain stressful conditions on neuronal cells such as aging and polyglutamine neurotoxicity. Cells stably expressing Ataxin-3 upregulate the mRNA levels of inflammatory response proteins, suggesting that inflammatory processes are involved in the pathogenesis of spinocerebellar ataxia type 3. Ataxin-3 binds to the N-terminus of two human homologs of the yeast DNA repair protein RAD23, HHR23A and HHR23B, which are important for nucleotide excision repair.

## REFERENCES


1. Gispert, S., et al. 1993. Chromosomal assignment of the second locus for autosomal dominant cerebellar ataxia (SCA2) to chromosome 12q23-24.1 Nat. Genet. 4: 295-299.
2. Pujana, M.A., et al. 1999. Spinocerebellar ataxias in Spanish patients: genetic analysis of familial and sporadic cases. The Ataxia Study Group. Hum. Genet. 104: 516-522.
3. Perez, M.K., et al. 1999. Ataxin-3 with an altered conformation that exposes the polyglutamine domain is associated with the nuclear matrix. Hum. Mol. Genet. 8: 2377-2385.
4. Huynh, D.P., et al. 2000. Nuclear localization or inclusion body formation of Ataxin-2 re not necessary for SCA2 pathogenesis in mouse or human. Nat. Genet. 26: 44-50.
5. Fujigasaki, H., et al. 2000. Ataxin-3 is translocated into the nucleus for the formation of intranuclear inclusions in normal and Machado-Joseph disease brains. Exp. Neurol. 165: 248-256.
6. Wang, G., et al. 2000. Ataxin-3, the MJD1 gene product, interacts with the two human homologs of yeast DNA repair protein RAD23, HHR23A and HHR23B. Hum. Mol. Genet. 9: 1795-1803.
7. Takahashi, J., et al. 2001. Recruitment of nonexpanded polyglutamine proteins to intranuclear aggregates in neuronal intranuclear hyaline inclusion disease. J. Neuropathol. Exp. Neurol. 60: 369-376.


## CHROMOSOMAL LOCATION

Genetic locus: ATXN3 (human) mapping to 14q32.12.

## RESEARCH USE

For research use only, not for use in diagnostic procedures.

## SOURCE

Ataxin-3 (C-5) is a mouse monoclonal antibody raised against amino acids 176-228 mapping within an internal region of Ataxin-3 of human origin.

## PRODUCT

Each vial contains 200 µg $IgG_1$ kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

## APPLICATIONS

Ataxin-3 (C-5) is recommended for detection of Ataxin-3 of human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for Ataxin-3 siRNA (h): sc-40358, Ataxin-3 shRNA Plasmid (h): sc-40358-SH and Ataxin-3 shRNA (h) Lentiviral Particles: sc-40358-V.

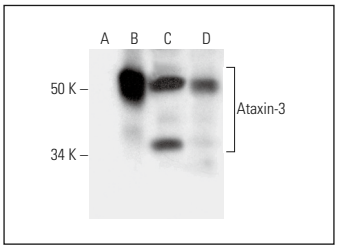
Molecular Weight of Ataxin-3: 42 kDa.

Positive Controls: Ataxin-3 (h): 293T Lysate: sc-114977, U-87 MG cell lysate: sc-2411 or human hippocampus tissue extract.

## RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

## DATA




Ataxin-3 (C-5): sc-393193. Western blot analysis of Ataxin-3 expression in non-transfected 293T: sc-117752 (**A**), human Ataxin-3 transfected 293T: sc-114977 (**B**) and U-87 MG (**C**) whole cell lysates and human hippocampus tissue extract (**D**).

## STORAGE

Store at 4° C, **DO NOT FREEZE**. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.