SANTA CRUZ BIOTECHNOLOGY, INC.

Ataxin-3 (A-7): sc-398114

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. 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.

CHROMOSOMAL LOCATION

Genetic locus: ATXN3 (human) mapping to 14q32.12; Atxn3 (mouse) mapping to 12 E.

SOURCE

Ataxin-3 (A-7) is a mouse monoclonal antibody raised against amino acids 176-234 mapping within an internal region of Ataxin-3 of mouse origin.

PRODUCT

Each vial contains 200 µg IgG_{2b} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Ataxin-3 (A-7) is available conjugated to agarose (sc-398114 AC), 500 µg/ 0.25 ml agarose in 1 ml, for IP; to HRP (sc-398114 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-398114 PE), fluorescein (sc-398114 FITC), Alexa Fluor® 488 (sc-398114 AF488), Alexa Fluor® 546 (sc-398114 AF546), Alexa Fluor® 594 (sc-398114 AF594) or Alexa Fluor® 647 (sc-398114 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-398114 AF680) or Alexa Fluor® 790 (sc-398114 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

RESEARCH USE

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

APPLICATIONS

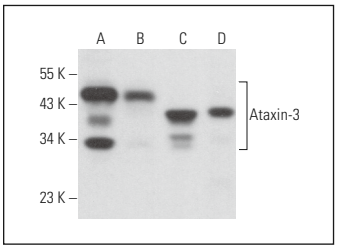
Ataxin-3 (A-7) is recommended for detection of Ataxin-3 of mouse, rat and human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500), immunohistochemistry (including paraffin-embedded sections) (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 siRNA (m): sc-40359, Ataxin-3 shRNA Plasmid (h): sc-40358-SH, Ataxin-3 shRNA Plasmid (m): sc-40359-SH, Ataxin-3 shRNA (h) Lentiviral Particles: sc-40358-V and Ataxin-3 shRNA (m) Lentiviral Particles: sc-40359-V.

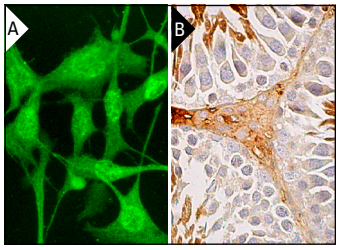
Molecular Weight of Ataxin-3: 42 kDa.

Positive Controls: CCRF-CEM cell lysate: sc-2225, Caki-1 cell lysate: sc-2224 or NIH/3T3 whole cell lysate: sc-2210.

DATA



Ataxin-3 (A-7): sc-398114. Western blot analysis of Ataxin-3 expression in CCRF-CEM (**A**), Caki-1 (**B**), NIH/3T3 (**C**) and SP2/0 (**D**) whole cell lysates.



Ataxin-3 (A-7): sc-398114. Immunofluorescence staining of methanol-fixed NIH/3T3 cells showing nuclear and cytoplasmic localization (**A**). Immunoperoxidase staining of formalin fixed, paraffin-embedded rat testis tissue showing nuclear and cytoplasmic staining of subset of cells in seminiferous ducts and cytoplasmic staining of Leydig cells (**B**).

SELECT PRODUCT CITATIONS

1. Tong, X., et al. 2021. Identification of a druggable protein-protein interaction site between mutant p53 and its stabilizing chaperone DNAJA1. J. Biol. Chem. 296: 100098.
2. Wu, X., et al. 2023. Involvement of Ataxin-3 (ATXN3) in the malignant progression of pancreatic cancer via deubiquitinating HDAC6. Pancreatology 23: 630-641.

STORAGE

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

PROTOCOLS

See our web site at www.scbt.com for detailed protocols and support products.