SANTA CRUZ BIOTECHNOLOGY, INC.

Ataxin-3 (h): 293T Lysate: sc-114977

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

Autosomal dominant cerebellar ataxias are a group of neurodegenerative 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 poly-glutamine 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 are not necessary for SCA2 pathogenesis in mouse or human. Nat. Genet. 26: 44-50.

CHROMOSOMAL LOCATION

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

PRODUCT

Ataxin-3 (h): 293T Lysate represents a lysate of human Ataxin-3 transfected 293T cells and is provided as 100 µg protein in 200 µl SDS-PAGE buffer.

STORAGE

Store at -20° C. Repeated freezing and thawing should be minimized. Sample vial should be boiled once prior to use. Non-hazardous. No MSDS required.

PROTOCOLS

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

APPLICATIONS

Ataxin-3 (h): 293T Lysate is suitable as a Western Blotting positive control for human reactive Ataxin-3 antibodies. Recommended use: 10-20 µl per lane.

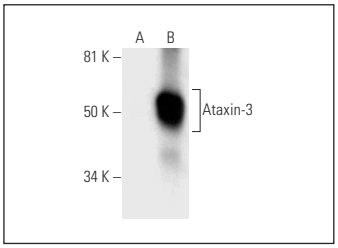
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

Ataxin-3 (C-5): sc-393193 is recommended as a positive control antibody for Western Blot analysis of enhanced human Ataxin-3 expression in Ataxin-3 transfected 293T cells (starting dilution 1:100, dilution range 1:100-1:1,000).

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.

DATA



Ataxin-3 (C-5): sc-393193. Western blot analysis of Ataxin-3 expression in non-transfected: sc-117752 (**A**) and human Ataxin-3 transfected: sc-114977 (**B**) 293T whole cell lysates.

RESEARCH USE

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