

Ataxin-3 (E-14): sc-18483

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.

CHROMOSOMAL LOCATION

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

SOURCE

Ataxin-3 (E-14) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the C-terminus of Ataxin-3 of rat origin.

PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-18483 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

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 or our catalog for detailed protocols and support products.

APPLICATIONS

Ataxin-3 (E-14) is recommended for detection of Ataxin-3 of rat, and, to a lesser extent, human origin by Western Blotting (starting dilution 1:200, 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.

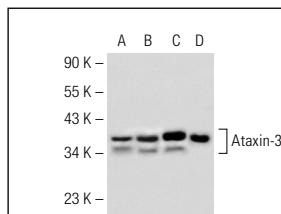
Molecular Weight of Ataxin-3: 42 kDa.

Positive Controls: KNRK nuclear extract: sc-2141 or 3611-RF nuclear extract: sc-2143.

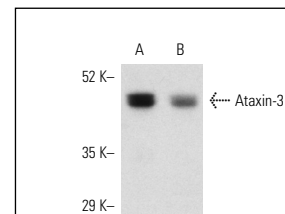
RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 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 donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

DATA



Ataxin-3 (E-14): sc-18483. Western blot analysis of Ataxin-3 expression in HeLa nuclear extract (A) and T98G (B), Caki-1 (C) and DU 145 (D) whole cell lysates.



Ataxin-3 (E-14): sc-18483. Western blot analysis of Ataxin-3 expression in 3611-RF (A) and KNRK (B) nuclear extracts.

RESEARCH USE

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

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Try **Ataxin-3 (A-7): sc-398114** or **Ataxin-3 (C-5): sc-393193**, our highly recommended monoclonal alternatives to Ataxin-3 (E-14).