

Ataxin-3 (N-20): sc-18480

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.

CHROMOSOMAL LOCATION

Genetic locus: ATXN3 (human) mapping to 14q32.12, ATXN3L (human) mapping to Xp22.2; Atxn3 (mouse) mapping to 12 E.

SOURCE

Ataxin-3 (N-20) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the N-terminus of Ataxin-3 of human 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-18480 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

Ataxin-3 (N-20) is recommended for detection of Ataxin-3 isoform 1 and ATXN3L of human origin and Ataxin-3 isoform 1 of mouse and rat 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).

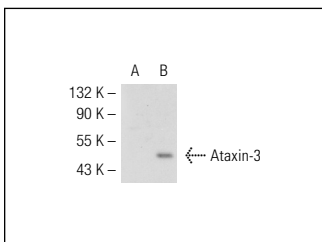
Ataxin-3 (N-20) is also recommended for detection of Ataxin-3 isoform 1 and ATXN3L in additional species, including canine, bovine, porcine and avian.

Suitable for use as control antibody for Ataxin-3 siRNA (m): sc-40359, Ataxin-3 shRNA Plasmid (m): sc-40359-SH and Ataxin-3 shRNA (m) Lentiviral Particles: sc-40359-V.

Molecular Weight of Ataxin-3: 42 kDa.

Positive Controls: Ataxin-3 (h): 293T Lysate: sc-114977, Caki-1 cell lysate: sc-2224 or DU 145 cell lysate: sc-2268.

DATA



Ataxin-3 (N-20): sc-18480. 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.

STORAGE

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

RESEARCH USE

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

PROTOCOLS

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



Try **Ataxin-3 (A-7): sc-398114** or **Ataxin-3 (C-5): sc-393193**, our highly recommended monoclonal alternatives to Ataxin-3 (N-20).