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

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

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

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 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 in additional species, including canine, bovine, porcine and avian.

Molecular Weight of Ataxin-3: 42 kDa.

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.

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.