

Ataxin-1 (H-21): sc-12527

BACKGROUND

Ataxin-1, also designated spinocerebellar ataxia type 1 protein (Sca-1), is differentially expressed and localizes to both the cytoplasm and the nucleus. Mutations in Ataxin-1 are associated with the onset of the autosomal dominant neurodegenerative disorder spinocerebellar ataxia type 1 (SCA-1), which is characterized by progressive neuronal loss in the cerebellum, muscle wasting and ataxia. In Purkinje cells, where SCA-1 is predominantly observed, Ataxin-1 has been shown to directly associate with the purkinje-enriched leucine-rich acidic nuclear protein (LANP) and the nuclear matrix-associated protein promyelocytic leukemia protein PML. In SCA-1, Ataxin-1 is mutated to encode a polyglutamine protein that forms nuclear aggregates, which interact significantly more strongly with LANP and contribute to the pathogenesis of SCA-1.

REFERENCES

1. Banfi, S., et al. 1994. Identification and characterization of the gene causing type 1 spinocerebellar ataxia. *Nat. Genet.* 7: 513-520.
2. Burright, E.N., et al. 1995. SCA1 transgenic mice: a model for neurodegeneration caused by an expanded CAG trinucleotide repeat. *Cell* 82: 937-948.

CHROMOSOMAL LOCATION

Genetic locus: ATXN1 (human) mapping to 6p22.3.

SOURCE

Ataxin-1 (H-21) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the N-terminus of Ataxin-1 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-12527 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

RESEARCH USE

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

APPLICATIONS

Ataxin-1 (H-21) is recommended for detection of Ataxin-1 of human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), 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).

Ataxin-1 (H-21) is also recommended for detection of Ataxin-1 in additional species, including equine and canine.

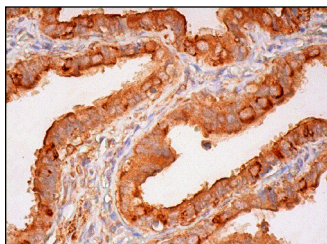
Suitable for use as control antibody for Ataxin-1 siRNA (h): sc-43624, Ataxin-1 shRNA Plasmid (h): sc-43624-SH and Ataxin-1 shRNA (h) Lentiviral Particles: sc-43624-V.

Molecular Weight of ATXN1: 98 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) 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. 3) Immunohistochemistry: use ImmunoCruz™: sc-2053 or ABC: sc-2023 goat IgG Staining Systems.

DATA



Ataxin-1 (H-21): sc-12527. Immunoperoxidase staining of formalin fixed, paraffin-embedded human fallopian tube tissue showing cytoplasmic staining of glandular cells.

SELECT PRODUCT CITATIONS

1. Hoshino, M., et al. 2004. General transcriptional repression by polyglutamine disease proteins is not directly linked to the presence of inclusion bodies. *Biochem. Biophys. Res. Commun.* 313: 110-116.
2. Tamura, T., et al. 2009. Glial cell lineage expression of mutant Ataxin-1 and Huntingtin induces developmental and late-onset neuronal pathologies in *Drosophila* models. *PLoS ONE* 4: e4262.

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.

MONOS
Satisfaction
Guaranteed

Try **Ataxin-1 (E-4): sc-514953** or **Ataxin-1 (B-3): sc-365343**, our highly recommended monoclonal alternatives to Ataxin-1 (H-21).