

Ataxin-1 (L-19): sc-12526

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.
3. Burright, E.N., et al. 1997. Identification of a self-association region within the SCA1 gene product, ataxin-1. *Hum. Mol. Genet.* 6: 513-518.
4. Skinner, P.J., et al. 1997. Ataxin-1 with an expanded glutamine tract alters nuclear matrix-associated structures. *Nature* 389: 971-974.
5. Matilla, A., et al. 1997. The cerebellar leucine-rich acidic nuclear protein interacts with ataxin-1. *Nature* 389: 974-978.
6. Klement, I.A., et al. 1998. Ataxin-1 nuclear localization and aggregation: role in polyglutamine-induced disease in SCA1 transgenic mice. *Cell* 95: 41-53.

CHROMOSOMAL LOCATION

Genetic locus: ATXN1 (human) mapping to 6p22.3; Atxn1 (mouse) mapping to 13 A5.

SOURCE

Ataxin-1 (L-19) 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-12526 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.

APPLICATIONS

Ataxin-1 (L-19) is recommended for detection of Ataxin-1 of human, and, to a lesser extent mouse 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).

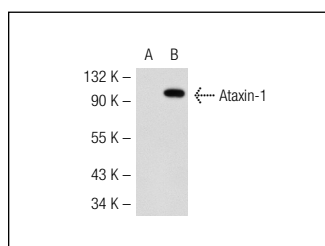
Suitable for use as control antibody for Ataxin-1 siRNA (h): sc-43624, Ataxin-1 siRNA (m): sc-40355, Ataxin-1 shRNA Plasmid (h): sc-43624-SH, Ataxin-1 shRNA Plasmid (m): sc-40355-SH, Ataxin-1 shRNA (h) Lentiviral Particles: sc-43624-V and Ataxin-1 shRNA Plasmid (m): sc-40355-SH.

Molecular Weight of Ataxin-1: 98 kDa.

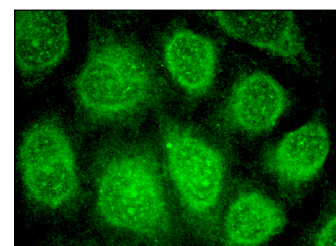
RECOMMENDED SECONDARY REAGENTS

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) 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-1 (L-19): sc-12526. Western blot analysis of Ataxin-1 expression in non-transfected: sc-117752 (A) and mouse Ataxin-1 transfected: sc-118599 (B) 293T whole cell lysates.



Ataxin-1 (L-19): sc-12526. Immunofluorescence staining of methanol-fixed HeLa cells showing nuclear localization.

RESEARCH USE

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

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 (L-19).