SANTA CRUZ BIOTECHNOLOGY, INC.

Ataxin-1 (E-4): sc-514953

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The Power to Question

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. SCA-1 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 (E-4) is a mouse monoclonal antibody raised against amino acids 578-752 mapping within an internal region of Ataxin-1 of human origin.

PRODUCT

Each vial contains 200 µg IgG_1 kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Ataxin-1 (E-4) is available conjugated to agarose (sc-514953 AC), 500 µg/ 0.25 ml agarose in 1 ml, for IP; to HRP (sc-514953 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-514953 PE), fluorescein (sc-514953 FITC), Alexa Fluor® 488 (sc-514953 AF488), Alexa Fluor® 546 (sc-514953 AF546), Alexa Fluor® 594 (sc-514953 AF594) or Alexa Fluor® 647 (sc-514953 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-514953 AF680) or Alexa Fluor® 790 (sc-514953 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

APPLICATIONS

Ataxin-1 (E-4) is recommended for detection of Ataxin-1 of mouse, rat and human origin by Western Blotting (starting dilution 1:100, 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 (m) Lentiviral Particles: sc-40355-V.

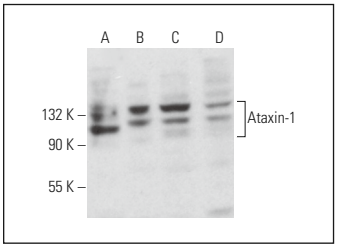
Molecular Weight of Ataxin-1: 98 kDa.

Positive Controls: SK-BR-3 cell lysate: sc-2218, NIH/3T3 whole cell lysate: sc-2210 or L6 whole cell lysate: sc-364196.

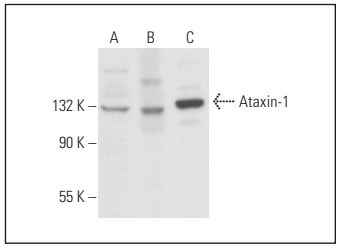
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 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 m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

DATA



Ataxin-1 (E-4): sc-514953. Western blot analysis of Ataxin-1 expression in NIH/3T3 (**A**), Sol8 (**B**), C2C12 (**C**) and A-10 (**D**) whole cell lysates.



Ataxin-1 (E-4): sc-514953. Western blot analysis of Ataxin-1 expression in SK-BR-3 (**A**), NIH/3T3 (**B**) and L6 (**C**) whole cell lysates.

SELECT PRODUCT CITATIONS

1. Yin, S., et al. 2021. Ccdc134 deficiency impairs cerebellar development and motor coordination. Genes Brain Behav. 20: e12763.

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.