Ataxin-1 (m): 293T Lysate: sc-118599



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

- Banfi, S., Servadio, A., Chung, M.Y., Kwiatkowski, T.J., Jr., McCall, A.E., Duvick, L.A., Shen, Y., Roth, E.J., Orr, H.T. and Zoghbi, H.Y. 1994. Identification and characterization of the gene causing type 1 spinocerebellar ataxia. Nat. Genet. 7: 513-520.
- Burright, E.N., Clark, H.B., Servadio, A., Matilla, T., Feddersen, R.M., Yunis, W.S., Duvick, L.A., Zoghbi, H.Y. and Orr, H.T. 1995. SCA-1 transgenic mice: a model for neurodegeneration caused by an expanded CAG trinucleotide repeat. Cell 82: 937-948.
- 3. Burright, E.N., Davidson, J.D., Duvick, L.A., Koshy, B., Zoghbi, H.Y. and Orr, H.T. 1997. Identification of a self-association region within the SCA1 gene product, Ataxin-1. Hum. Mol. Genet. 6: 513-518.
- Skinner, P.J., Koshy, B.T., Cummings, C.J., Klement, I.A., Helin, K., Servadio, A., Zoghbi, H.Y. and Orr, H.T. 1997. Ataxin-1 with an expanded glutamine tract alters nuclear matrix-associated structures. Nature 389: 971-974.
- Matilla, A., Koshy, B.T., Cummings, C.J., Isobe, T., Orr, H.T. and Zoghbi, H.Y. 1997. The cerebellar leucine-rich acidic nuclear protein interacts with Ataxin-1. Nature 389: 974-978.
- Klement, I.A., Skinner, P.J., Kaytor, M.D., Yi, H., Hersch, S.M., Clark, H.B., Zoghbi, H.Y. and Orr, H.T. 1998. Ataxin-1 nuclear localization and aggregation: role in polyglutamine-induced disease in SCA1 transgenic mice. Cell 95: 41-53.

CHROMOSOMAL LOCATION

Genetic locus: Sca1 (mouse) mapping to 13 A5.

PRODUCT

Ataxin-1 (m): 293T Lysate represents a lysate of mouse Ataxin-1 transfected 293T cells and is provided as 100 μ g protein in 200 μ l SDS-PAGE buffer.

STORAGE

Store at -20 $^{\circ}$ C. Repeated freezing and thawing should be minimized. Sample vial should be boiled once prior to use. Non-hazardous. No MSDS required.

RESEARCH USE

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

APPLICATIONS

Ataxin-1 (m): 293T Lysate is suitable as a Western Blotting positive control for mouse reactive Ataxin-1 antibodies. Recommended use: 10-20 μ l per lane

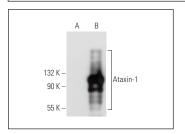
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

Ataxin-1 (E-4): sc-514953 is recommended as a positive control antibody for Western Blot analysis of enhanced mouse Ataxin-1 expression in Ataxin-1 transfected 293T cells (starting dilution 1:100, dilution range 1:100-1:1,000).

RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-lgG κ BP-HRP: sc-516102 or m-lgG κ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz MarkerTM Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048.

DATA



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

PROTOCOLS

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