

Ataxin-1 siRNA (h): sc-43624

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.

PRODUCT

Ataxin-1 siRNA (h) is a pool of 3 target-specific 19-25 nt siRNAs designed to knock down gene expression. Each vial contains 3.3 nmol of lyophilized siRNA, sufficient for a 10 μ M solution once resuspended using protocol below. Suitable for 50-100 transfections. Also see Ataxin-1 shRNA Plasmid (h): sc-43624-SH and Ataxin-1 shRNA (h) Lentiviral Particles: sc-43624-V as alternate gene silencing products.

For independent verification of Ataxin-1 (h) gene silencing results, we also provide the individual siRNA duplex components. Each is available as 3.3 nmol of lyophilized siRNA. These include: sc-43624A, sc-43624B and sc-43624C.

PROTOCOLS

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

STORAGE AND RESUSPENSION

Store lyophilized siRNA duplex at -20° C with desiccant. Stable for at least one year from the date of shipment. Once resuspended, store at -20° C, avoid contact with RNases and repeated freeze thaw cycles.

Resuspend lyophilized siRNA duplex in 330 μ l of the RNase-free water provided. Resuspension of the siRNA duplex in 330 μ l of RNase-free water makes a 10 μ M solution in a 10 μ M Tris-HCl, pH 8.0, 20 mM NaCl, 1 mM EDTA buffered solution.

APPLICATIONS

Ataxin-1 siRNA (h) is recommended for the inhibition of Ataxin-1 expression in human cells.

SUPPORT REAGENTS

For optimal siRNA transfection efficiency, Santa Cruz Biotechnology's siRNA Transfection Reagent: sc-29528 (0.3 ml), siRNA Transfection Medium: sc-36868 (20 ml) and siRNA Dilution Buffer: sc-29527 (1.5 ml) are recommended. Control siRNAs or Fluorescein Conjugated Control siRNAs are available as 10 μ M in 66 μ l. Each contain a scrambled sequence that will not lead to the specific degradation of any known cellular mRNA. Fluorescein Conjugated Control siRNAs include: sc-36869, sc-44239, sc-44240 and sc-44241. Control siRNAs include: sc-37007, sc-44230, sc-44231, sc-44232, sc-44233, sc-44234, sc-44235, sc-44236, sc-44237 and sc-44238.

GENE EXPRESSION MONITORING

Ataxin-1 (E-4): sc-514953 is recommended as a control antibody for monitoring of Ataxin-1 gene expression knockdown by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000) or immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500).

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgG κ BP-HRP: sc-516102 or m-IgG κ 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) Immunofluorescence: use m-IgG κ BP-FITC: sc-516140 or m-IgG κ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

RT-PCR REAGENTS

Semi-quantitative RT-PCR may be performed to monitor Ataxin-1 gene expression knockdown using RT-PCR Primer: Ataxin-1 (h)-PR: sc-43624-PR (20 μ l). Annealing temperature for the primers should be 55-60° C and the extension temperature should be 68-72° C.

RESEARCH USE

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