Ataxin-7 siRNA (h): sc-40360



The Power to Question

BACKGROUND

The human Ataxin-7 gene, also known as spinocerebellar ataxia 7 or SCA7, maps to chromosome 3p14.1, has a 2,727-bp open reading frame, and encodes a 892 amino acid protein containing a nuclear localization signal and a polyglutamine tract. SCA7 is an autosomal dominant neurodegenerative disorder characterized by ataxia and selective neuronal cell loss caused by the expansion of a translated CAG repeat encoding a polyglutamine tract in Ataxin-7, which is the SCA7 gene product. Ataxin-7 is expressed within neurons both affected and unaffected in SCA7 pathology with subcellular localization being variable depending upon the neuronal subtype. Polyglutamine expanded in Ataxin-7 may carry out its pathogenic effects in the nucleus by altering the matrix-associated nuclear structure and/or by disrupting nucleolar function.

REFERENCES

- David, G., Abbas, N., Stevanin, G., Durr, A., Yvert, G., Cancel, G., Weber, C., Imbert, G., Saudou, F., Mandel, J.L. and Brice, A. 1997. Cloning of the SCA7 gene reveals a highly unstable CAG repeat expansion. Nat. Genet. 17: 65-70.
- 2. Kaytor, M.D., Duvick, L.A., Skinner, P.J., Koob, M.D., Ranum, L.P. and Orr, H.T. 1999. Nuclear localization of the spinocerebellar ataxia type 7 protein, Ataxin-7. Hum. Mol. Genet. 8: 1657-1664.
- 3. La Spada, A.R., Fu, Y.H., Sopher, B.L., Libby, R.T., Wang, X., Li, L.Y., Einum, D.D., Huang, J., Possin, D.E., Smith, A.C., Martinez, R.A., Koszdin, K.L., Treuting, P.M., Ware, C.B., Hurley, J.B., et al. 2001. Polyglutamine-expanded Ataxin-7 antagonizes CRX function and induces cone-rod dystrophy in a mouse model of SCA7. Neuron 31: 913-927.
- 4. Matilla, A., Gorbea, C., Einum, D.D., Townsend, J., Michalik, A., van Broeckhoven, C., Jensen, C.C., Murphy, K.J., Ptacek, L.J. and Fu, Y.H. 2001. Association of Ataxin-7 with the proteasome subunit S4 of the 19S regulatory complex. Hum. Mol. Genet. 10: 2821-2831.
- 5. Einum, D.D., Townsend, J.J., Ptacek, L.J. and Fu, Y.H. 2001. Ataxin-7 expression analysis in controls and spinocerebellar ataxia type 7 patients. Neurogenetics 3: 83-90.
- Online Mendelian Inheritance in Man, OMIM™. 2001. Johns Hopkins University, Baltimore, MD. MIM Number: 164500. World Wide Web URL: http://www.ncbi.nlm.nih.gov/omim/

CHROMOSOMAL LOCATION

Genetic locus: ATXN7 (human) mapping to 3p14.1.

RESEARCH USE

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

PROTOCOLS

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

PRODUCT

Ataxin-7 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-7 shRNA Plasmid (h): sc-40360-SH and Ataxin-7 shRNA (h) Lentiviral Particles: sc-40360-V as alternate gene silencing products.

For independent verification of Ataxin-7 (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-40360A, sc-40360B and sc-40360C.

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-7 siRNA (h) is recommended for the inhibition of Ataxin-7 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 µM in 66 µl. 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.

RT-PCR REAGENTS

Semi-quantitative RT-PCR may be performed to monitor Ataxin-7 gene expression knockdown using RT-PCR Primer: Ataxin-7 (h)-PR: sc-40360-PR (20 μ l). Annealing temperature for the primers should be 55-60° C and the extension temperature should be 68-72° C.

Santa Cruz Biotechnology, Inc. 1.800.457.3801 831.457.3801 fax 831.457.3801 Europe +00800 4573 8000 49 6221 4503 0 www.scbt.com