

Wolframin siRNA (h): sc-61804

BACKGROUND

The Wolframin gene encodes a protein found in endoplasmic reticulum membrane of several tissues including brain, pancreas, lung and placenta. Loss-of-function mutations in both alleles result in Wolfram syndrome (also known as DIDMOAD, an autosomal recessive disorder that causes juvenile diabetes mellitus, diabetes insipidus, optic atrophy and a number of neurological symptoms including deafness, ataxia and peripheral neuropathy. A large number and variety of mutations in this gene, particularly in exon 8, can be associated with Wolfram syndrome. Mutations in this gene can also cause autosomal dominant deafness 6 (DFNA6), also known as DFNA14 or DFNA38.

REFERENCES

1. Osman, A.A., et al. 2003. Wolframin expression induces novel ion channel activity in endoplasmic reticulum membranes and increases intracellular calcium. *J. Biol. Chem.* 278: 52755-52762.
2. Larsen, Z.M., et al. 2004. Evidence for linkage on chromosome 4p16.1 in type 1 diabetes Danish families and complete mutation scanning of the WFS1 (Wolframin) gene. *Diabet. Med.* 21: 218-222.
3. Smith, C.J., et al. 2004. Phenotype-genotype correlations in a series of Wolfram syndrome families. *Diabetes Care* 27: 2003-2009.
4. Yamaguchi, S., et al. 2004. Endoplasmic reticulum stress and N-glycosylation modulate protein. *Biochem. Biophys. Res. Commun.* 325: 250-256.
5. Fonseca, S.G., et al. 2005. WFS1 is a novel component of the unfolded protein response and maintains homeostasis of the endoplasmic reticulum in pancreatic β -cells. *J. Biol. Chem.* 280: 39609-39615.
6. Koido, K., et al. 2005. Polymorphisms in Wolframin (WFS1) gene are possibly related to mood disorders. *Int. J. Neuropsychopharmacol.* 8: 235-244.
7. Philbrook, C., et al. 2005. Expressional and functional studies of Wolframin, the gene function deficient in Wolfram syndrome, in mice and patient cells. *Exp. Gerontol.* 40: 671-678.
8. Swift, M. and Swift, R.G. 2005. Wolframin mutations and hospitalization for psychiatric illness. *Mol. Psychiatry* 10: 799-803.
9. Domenech, E., et al. 2006. Wolfram/DIDMOAD syndrome, a heterogenic and molecularly complex neurodegenerative disease. *Pediatr. Endocrinol. Rev.* 3: 249-257.

CHROMOSOMAL LOCATION

Genetic locus: WFS1 (human) mapping to 4p16.1.

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

Wolframin siRNA (h) is a target-specific 19-25 nt siRNA 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 Wolframin shRNA Plasmid (h): sc-61804-SH and Wolframin shRNA (h) Lentiviral Particles: sc-61804-V as alternate gene silencing products.

For independent verification of Wolframin (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-61804A, sc-61804B and sc-61804C.

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

Wolframin siRNA (h) is recommended for the inhibition of Wolframin 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.

RT-PCR REAGENTS

Semi-quantitative RT-PCR may be performed to monitor Wolframin gene expression knockdown using RT-PCR Primer: Wolframin (h)-PR: sc-61804-PR (20 μ l). Annealing temperature for the primers should be 55-60° C and the extension temperature should be 68-72° C.