

Retinoschisin siRNA (h): sc-44771

BACKGROUND

X-linked juvenile retinoschisis (XLRS), the most common form of early onset macular degeneration in males, is characterized by delamination of the inner retinal layers and severe loss of vision. XLRS is caused by over 125 different mutations in the RS1 gene, which encodes the discoidin domain-containing protein Retinoschisin. Retinoschisin functions as a cell adhesion protein that maintains the cellular organization and synaptic structure of the retina. It is secreted from retinal tissues, specifically photoreceptor and bipolar cells, as an octamer, the subunits of which are joined together by Cys 59-Cys 223 intermolecular disulfide bonds. The interaction of cysteine residues in the Retinoschisin protein are critical for proper folding and subunit assembly. Misfolding of the discoidin domain, defective disulfide-linked subunit assembly and inability of Retinoschisin to insert into the endoplasmic reticulum membrane are responsible for the loss of function of Retinoschisin and the pathogenesis of XLRS.

REFERENCES

1. Grayson, C., et al. 2000. Retinoschisin, the X-linked retinoschisis protein, is a secreted photoreceptor protein and is expressed and released by Veri-Rb1 cells. *Hum. Mol. Genet.* 9: 1873-1879.
2. Molday, L.L., et al. 2001. Expression of X-linked retinoschisis protein RS1 in photoreceptor and bipolar cells. *Invest. Ophthalmol. Vis. Sci.* 42: 816-825.
3. Wu, W.W., et al. 2003. Defective discoidin domain structure, subunit assembly and endoplasmic reticulum processing of Retinoschisin are primary mechanisms responsible for X-linked retinoschisis. *J. Biol. Chem.* 278: 28139-28146.
4. Reid, S.N., et al. 2003. Retinoschisin, a photoreceptor-secreted protein, and its interaction with bipolar and muller cells. *J. Neurosci.* 23: 6030-6040.
5. Takada, Y., et al. 2004. A retinal neuronal developmental wave of Retinoschisin expression begins in ganglion cells during layer formation. *Invest. Ophthalmol. Vis. Sci.* 45: 3302-3312.
6. Tantri, A., et al. 2004. X-linked retinoschisis: a clinical and molecular genetic review. *Surv. Ophthalmol.* 49: 214-230.

CHROMOSOMAL LOCATION

Genetic locus: RS1 (human) mapping to Xp22.13.

PRODUCT

Retinoschisin 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 Retinoschisin shRNA Plasmid (h): sc-44771-SH and Retinoschisin shRNA (h) Lentiviral Particles: sc-44771-V as alternate gene silencing products.

For independent verification of Retinoschisin (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-44771A, sc-44771B and sc-44771C.

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

Retinoschisin siRNA (h) is recommended for the inhibition of Retinoschisin 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 Retinoschisin gene expression knockdown using RT-PCR Primer: Retinoschisin (h)-PR: sc-44771-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.

PROTOCOLS

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