

Harmonin siRNA (h): sc-40648

BACKGROUND

The gene encoding human harmonin (NY-CO-38/PDZ-73) maps to chromosome 11p15.1. Mutations in the harmonin gene cause Usher syndrome type I subtype C and non-syndromic deafness. Alternative splicing generates three harmonin isoforms. Harmonin isoform 1 contains three PDZ protein-protein interaction domains. Renal and colon cancer patients frequently develop autoantibodies to harmonin, which is present in kidney, brain, small intestine and colon. Sensory hair cells (stereocilia) in the inner ear also express harmonin. The first PDZ domain of Harmonin binds mutated in colon cancer 2 (MCC2) at the carboxy terminal. Harmonin also interacts with cadherin 23 and myosin VIIA in growing stereocilia of the inner ear. The harmonin/cadherin 23/myosin VIIA complex influences the shaping of a functional stereocilia bundle.

REFERENCES

1. Scanlan, M.J., et al. 1999. Isoforms of the human PDZ-73 protein exhibit differential tissue expression. *Biochim. Biophys. Acta* 1445: 39-52.
2. Verpy, E., et al. 2000. A defect in harmonin, a PDZ domain-containing protein expressed in the inner ear sensory hair cells, underlies Usher syndrome type 1C. *Nat. Genet.* 26: 51-55.
3. Bitner-Glindzic, M., et al. 2000. A recessive contiguous gene deletion causing infantile hyperinsulinism, enteropathy and deafness identifies the Usher type 1C gene. *Nat. Genet.* 26: 56-60.
4. Ishikawa, S., et al. 2001. Interaction of MCC2, a novel homologue of MCC tumor suppressor, with PDZ-domain Protein AIE-75. *Gene* 267: 101-110.
5. Ahmed, Z.M., et al. 2002. Nonsyndromic recessive deafness DFNB18 and Usher syndrome type IC are allelic mutations of USH1C. *Hum. Genet.* 110: 527-531.
6. Ouyang, X.M., et al. 2002. Mutations in the alternatively spliced exons of USH1C cause non-syndromic recessive deafness. *Hum. Genet.* 111: 26-30.
7. Siemens, J., et al. 2002. The Usher syndrome proteins cadherin 23 and harmonin form a complex by means of PDZ-domain interactions. *Proc. Natl. Acad. Sci. USA* 99: 14946-14951.
8. Boeda, B., et al. 2002. Myosin VIIa, harmonin and cadherin 23, three Usher I gene products that cooperate to shape the sensory hair cell bundle. *EMBO J.* 21: 6689-6699.

CHROMOSOMAL LOCATION

Genetic locus: USH1C (human) mapping to 11p15.1.

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

Harmonin 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 Harmonin shRNA Plasmid (h): sc-40648-SH and Harmonin shRNA (h) Lentiviral Particles: sc-40648-V as alternate gene silencing products.

For independent verification of Harmonin (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-40648A, sc-40648B and sc-40648C.

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

Harmonin siRNA (h) is recommended for the inhibition of Harmonin 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 Harmonin gene expression knockdown using RT-PCR Primer: Harmonin (h)-PR: sc-40648-PR (20 μ l). Annealing temperature for the primers should be 55-60° C and the extension temperature should be 68-72° C.