

USH1G siRNA (h): sc-63189

BACKGROUND

USH1G (Usher syndrome 1G), also known as ANKS4A or SANS, is a 461 amino acid protein that contains three ANK repeats and one SAM (sterile α motif) domain. Expressed in the small intestine, as well as in tissue of the eye and inner ear, USH1G associates with Harmonin and is thought to play a role in the development and maintenance of both auditory and visual systems, specifically by mediating the cohesion of hair bundles formed by inner ear sensory cells. Defects in the gene encoding USH1G are the cause of Usher syndrome type 1G (USH1G), a heterogeneous condition that is characterized by profound congenital sensorineural deafness, absent vestibular function and prepubertal onset of progressive retinitis pigmentosa, ultimately leading to blindness.

REFERENCES

1. Kitamura, K., et al. 1992. Ultrastructural findings in the inner ear of Jackson shaker mice. *Acta Otolaryngol.* 112: 622-627.
2. Kikkawa, Y., et al. 2003. Mutations in a new scaffold protein Sans cause deafness in Jackson shaker mice. *Hum. Mol. Genet.* 12: 453-461.
3. Weil, D., et al. 2003. Usher syndrome type I G (USH1G) is caused by mutations in the gene encoding SANS, a protein that associates with the USH1C protein, harmonin. *Hum. Mol. Genet.* 12: 463-471.
4. Ouyang, X.M., et al. 2005. Characterization of Usher syndrome type I gene mutations in an Usher syndrome patient population. *Hum. Genet.* 116: 292-299.
5. Adato, A., et al. 2005. Interactions in the network of Usher syndrome type 1 proteins. *Hum. Mol. Genet.* 14: 347-356.
6. Aller, E., et al. 2007. Screening of the USH1G gene among Spanish patients with Usher syndrome. Lack of mutations and evidence of a minor role in the pathogenesis of the syndrome. *Ophthalmic Genet.* 28: 151-155.
7. Online Mendelian Inheritance in Man, OMIM™. 2007. Johns Hopkins University, Baltimore, MD. MIM Number: 607696. World Wide Web URL: <http://www.ncbi.nlm.nih.gov/omim/>
8. Baux, D., et al. 2008. UMD-USHbases: a comprehensive set of databases to record and analyse pathogenic mutations and unclassified variants in seven Usher syndrome causing genes. *Hum. Mutat.* 29: E76-E87.

CHROMOSOMAL LOCATION

Genetic locus: USH1G (human) mapping to 17q25.1.

PRODUCT

USH1G 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 USH1G shRNA Plasmid (h): sc-63189-SH and USH1G shRNA (h) Lentiviral Particles: sc-63189-V as alternate gene silencing products.

For independent verification of USH1G (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-63189A, sc-63189B and sc-63189C.

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

USH1G siRNA (h) is recommended for the inhibition of USH1G 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

USH1G (D-10): sc-514418 is recommended as a control antibody for monitoring of USH1G 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 USH1G gene expression knockdown using RT-PCR Primer: USH1G (h)-PR: sc-63189-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.