

TMC1 siRNA (m): sc-154314

BACKGROUND

TMC1 (transmembrane channel-like protein 1) is a 760 amino acid multi-pass membrane protein that belongs to the TMC family. While it is expressed in fetal cochlea, TMC1 is also detected at low levels in placenta and testis. Required for the normal function of cochlear hair cells, mutations in the TMC1 gene lead to hearing impairment. Defects in TMC1 are the cause of deafness autosomal dominant type 36 (DFNA36), a form of sensorineural hearing loss. Sensorineural deafness results from damage to the neural receptors of the inner ear, the nerve pathways to the brain or the area of the brain that receives sound information. DFNA36 is a bilateral hearing loss, beginning at 5-10 years of age and progressing to profound deafness within 10-15 years. Defects in TMC1 are the cause of deafness autosomal recessive type 7 (DFNB7), also known as autosomal recessive neurosensory deafness type 11 (DFNB11). This hearing loss is congenital and profound.

REFERENCES

1. Online Mendelian Inheritance in Man, OMIM[™]. 2002. Johns Hopkins University, Baltimore, MD. MIM Number: 606706. World Wide Web URL: <http://www.ncbi.nlm.nih.gov/omim/>
2. Tlili, A., et al. 2008. TMC1 but not TMC2 is responsible for autosomal recessive nonsyndromic hearing impairment in Tunisian families. *Audiol. Neurotol.* 13: 213-218.
3. Hilgert, N., et al. 2008. Mutation analysis of TMC1 identifies four new mutations and suggests an additional deafness gene at loci DFNA36 and DFNB7/11. *Clin. Genet.* 74: 223-232.
4. Sirmaci, A., et al. 2009. Mutations in TMC1 contribute significantly to nonsyndromic autosomal recessive sensorineural hearing loss: a report of five novel mutations. *Int. J. Pediatr. Otorhinolaryngol.* 73: 699-705.
5. Hilgert, N., et al. 2009. Amino acid 572 in TMC1: hot spot or critical functional residue for dominant mutations causing hearing impairment. *J. Hum. Genet.* 54: 188-190.
6. Hildebrand, M.S., et al. 2010. Mutations in TMC1 are a common cause of DFNB7/11 hearing loss in the Iranian population. *Ann. Otol. Rhinol. Laryngol.* 119: 830-835.
7. Yang, T., et al. 2010. A novel mutation adjacent to the Bth mouse mutation in the TMC1 gene makes this mouse an excellent model of human deafness at the DFNA36 locus. *Clin. Genet.* 77: 395-398.
8. Ben Saïd, M., et al. 2010. High frequency of the p.R34X mutation in the TMC1 gene associated with nonsyndromic hearing loss is due to founder effects. *Genet. Test. Mol. Biomarkers* 14: 307-311.
9. de Heer, A.M., et al. 2011. Progressive sensorineural hearing loss and normal vestibular function in a Dutch DFNB7/11 family with a novel mutation in TMC1. *Audiol. Neurotol.* 16: 93-105.

CHROMOSOMAL LOCATION

Genetic locus: Tmc1 (mouse) mapping to 19 B.

RESEARCH USE

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

PRODUCT

TMC1 siRNA (m) 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 TMC1 shRNA Plasmid (m): sc-154314-SH and TMC1 shRNA (m) Lentiviral Particles: sc-154314-V as alternate gene silencing products.

PROTOCOLS

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

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

TMC1 siRNA (m) is recommended for the inhibition of TMC1 expression in mouse 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 TMC1 gene expression knockdown using RT-PCR Primer: TMC1 (m)-PR: sc-154314-PR (20 μ l). Annealing temperature for the primers should be 55-60° C and the extension temperature should be 68-72° C.