

TMEM126A siRNA (h): sc-96524

BACKGROUND

TMEM126A (transmembrane protein 126A) is a 195 amino acid multi-pass membrane protein that is encoded by a gene which maps to human chromosome 11 and may be involved in the pathogenesis of autosomal-recessive nonsyndromic optic atrophy. With approximately 135 million base pairs and 1,400 genes, chromosome 11 comprises approximately 4% of human genomic DNA and is considered a gene and disease association dense chromosome. The chromosome 11 encoded Atm gene is important for regulation of cell cycle arrest and apoptosis following double strand DNA breaks. Atm mutation leads to the disorder known as ataxia-telangiectasia. The blood disorders Sickle cell anemia and thalassemia are caused by HBB gene mutations, while Wilms' tumors, WAGR syndrome and Denys-Drash syndrome are associated with mutations of the WT1 gene. Jervell and Lange-Nielsen syndrome, Jacobsen syndrome, Niemann-Pick disease, hereditary angioedema and Smith-Lemli-Opitz syndrome are also associated with defects in chromosome 11-encoded genes.

REFERENCES

1. Yerushalmi, G.M., Leibowitz-Amit, R., Shaharabany, M. and Tsarfaty, I. 2002. Met-HGF/SF signal transduction induces mimp, a novel mitochondrial carrier homologue, which leads to mitochondrial depolarization. *Neoplasia* 4: 510-522.
2. Gross, A. 2005. Mitochondrial carrier homolog 2: a clue to cracking the Bcl-2 family riddle? *J. Bioenerg. Biomembr.* 37: 113-119.
3. Grinberg, M., Schwarz, M., Zaltsman, Y., Eini, T., Niv, H., Pietrokovski, S. and Gross, A. 2005. Mitochondrial carrier homolog 2 is a target of tBID in cells signaled to die by tumor necrosis factor alpha. *Mol. Cell. Biol.* 25: 4579-4590.
4. Schwarz, M., Andrade-Navarro, M.A. and Gross, A. 2007. Mitochondrial carriers and pores: key regulators of the mitochondrial apoptotic program? *Apoptosis* 12: 869-876.
5. Hanein, S., Perrault, I., Roche, O., Gerber, S., Khadom, N., Rio, M., Boddaert, N., Jean-Pierre, M., Brahimi, N., Serre, V., Chretien, D., Delphin, N., Fares-Taie, L., Lachheb, S., Rotig, A., Meire, F., Munnich, A., et al. 2009. TMEM126A, encoding a mitochondrial protein, is mutated in autosomal-recessive nonsyndromic optic atrophy. *Am. J. Hum. Genet.* 84: 493-498.
6. Meyer, E., Michaelides, M., Tee, L.J., Robson, A.G., Rahman, F., Pasha, S., Luxon, L.M., Moore, A.T. and Maher, E.R. 2010. Nonsense mutation in TMEM126A causing autosomal recessive optic atrophy and auditory neuropathy. *Mol. Vis.* 16: 650-664.
7. Desir, J., Coppieters, F., Van Regemorter, N., De Baere, E., Abramowicz, M. and Cordonnier, M. 2012. TMEM126A mutation in a Moroccan family with autosomal recessive optic atrophy. *Mol. Vis.* 18: 1849-1857.
8. Hanein, S., Garcia, M., Fares-Taie, L., Serre, V., De Keyser, Y., Delaveau, T., Perrault, I., Delphin, N., Gerber, S., Schmitt, A., Masse, J.M., Munnich, A., Kaplan, J., Devaux, F. and Rozet, J.M. 2013. TMEM126A is a mitochondrial located mRNA (MLR) protein of the mitochondrial inner membrane. *Biochim. Biophys. Acta* 1830: 3719-3733.

CHROMOSOMAL LOCATION

Genetic locus: TMEM126A (human) mapping to 11q14.1.

PRODUCT

TMEM126A siRNA (h) is a pool of 2 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 TMEM126A shRNA Plasmid (h): sc-96524-SH and TMEM126A shRNA (h) Lentiviral Particles: sc-96524-V as alternate gene silencing products.

For independent verification of TMEM126A (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-96524A and sc-96524B.

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

TMEM126A siRNA (h) is recommended for the inhibition of TMEM126A 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 TMEM126A gene expression knockdown using RT-PCR Primer: TMEM126A (h)-PR: sc-96524-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.