



RARS2 siRNA (h): sc-95647

BACKGROUND

RARS2, also known as probable arginyl-tRNA synthetase, is a 578 amino acid mitochondrial protein that belongs to the class-I aminoacyl-tRNA synthetase family. RARS2 mutations are predicted to result in a frameshift, skipping of exon 2, and abolished enzymatic activity. Defects in RARS2 are the cause of pontocerebellar hypoplasia type 6 (PCH6), also known as fatal infantile encephalopathy with mitochondrial respiratory chain defects. Pontocerebellar hypoplasia (PCH) is a heterogeneous group of disorders resulting in abnormal growth and function of brainstem and cerebellum. Comprising of 20 exons, the RARS2 gene is conserved in chimpanzee, canine, bovine, mouse, rat, chicken, zebrafish, fruit fly, mosquito, *S. pombe*, *S. cerevisiae*, *K. lactis*, *E. gossypii*, *M. grisea* and *N. crassa*, and maps to human chromosome 6q15.

REFERENCES

1. Mungall, A.J., et al. 2003. The DNA sequence and analysis of human chromosome 6. *Nature* 425: 805-811.
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3. Online Mendelian Inheritance in Man, OMIM™. 2007. Johns Hopkins University, Baltimore, MD. MIM Number: 611524. World Wide Web URL: <http://www.ncbi.nlm.nih.gov/omim/>
4. Pérez-Martínez, X., et al. 2008. Protein synthesis and assembly in mitochondrial disorders. *Curr. Top. Med. Chem.* 8: 1335-1350.
5. Rankin, J., et al. 2010. Pontocerebellar hypoplasia type 6: a British case with PEHO-like features. *Am. J. Med. Genet. A* 152A: 2079-2084.
6. Graham, J.M., et al. 2010. Molecular and neuroimaging findings in pontocerebellar hypoplasia type 2 (PCH2): is prenatal diagnosis possible? *Am. J. Med. Genet. A* 152A: 2268-2276.
7. Namavar, Y., et al. 2011. Clinical, neuroradiological and genetic findings in pontocerebellar hypoplasia. *Brain* 134: 143-156.

CHROMOSOMAL LOCATION

Genetic locus: RARS2 (human) mapping to 6q15.

PRODUCT

RARS2 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 RARS2 shRNA Plasmid (h): sc-95647-SH and RARS2 shRNA (h) Lentiviral Particles: sc-95647-V as alternate gene silencing products.

For independent verification of RARS2 (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-95647A, sc-95647B and sc-95647C.

RESEARCH USE

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

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

RARS2 siRNA (h) is recommended for the inhibition of RARS2 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 RARS2 gene expression knockdown using RT-PCR Primer: RARS2 (h)-PR: sc-95647-PR (20 μ l). Annealing temperature for the primers should be 55-60° C and the extension temperature should be 68-72° C.

PROTOCOLS

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