



WBSCR27 siRNA (h): sc-89703

BACKGROUND

Williams-Beuren syndrome (WBS) is a developmental disorder caused by the hemizygous microdeletion on chromosome 7q11.23. WBS is an autosomal dominant genetic condition that is characterized by physical, cognitive and behavioral traits. The physical traits associated with WBS include facial dysmorphism, vascular stenoses, growth deficiencies, dental anomalies and neurologic and musculoskeletal abnormalities. Mild retardation, a weakness in visual-spatial skills, anxiety and a short attention span are typical cognitive and behavioral traits of WBS patients. The WBSCR27 gene is located within the WBS deletion and may contribute to the developmental symptoms found in WBS because of a loss of the encoded transcription factor. WBSCR27 (Williams-Beuren syndrome chromosomal region 27 protein) is a 245 amino acid protein and is encoded by a gene located on human chromosome 7q11.23. Certain cardiovascular and musculo-skeletal abnormalities may be the result of haploinsufficiency of WBSCR27.

REFERENCES

1. Morris, C.A., Demsey, S.A., Leonard, C.O., Dilts, C. and Blackburn, B.L. 1988. Natural history of Williams syndrome: physical characteristics. *J. Pediatr.* 113: 318-326.
2. Lashkari, A., Smith, A.K. and Graham, J.M. 1999. Williams-Beuren syndrome: an update and review for the primary physician. *Clin. Pediatr.* 38: 189-208.
3. Bellugi, U., Lichtenberger, L., Mills, D., Galaburda, A. and Korenberg, J.R. 1999. Bridging cognition, the brain and molecular genetics: evidence from Williams syndrome. *Trends Neurosci.* 22: 197-207.
4. Hillier, L.W., Fulton, R.S., Fulton, L.A., Graves, T.A., Pepin, K.H., Wagner-McPherson, C., Layman, D., Maas, J., Jaeger, S., Walker, R., Wylie, K., et al. 2003. The DNA sequence of human chromosome 7. *Nature* 424: 157-164.
5. Gerhard, D.S., Wagner, L., Feingold, E.A., Shenmen, C.M., Grouse, L.H., Schuler, G., Klein, S.L., Old, S., Rasooly, R., Good, P., Guyer, M., Peck, A.M., Derge, J.G., et al. 2004. The status, quality, and expansion of the NIH full-length cDNA project: the Mammalian Gene Collection (MGC). *Genome Res.* 14: 2121-2127.
6. Micale, L., Fusco, C., Augello, B., Napolitano, L.M., Dermitzakis, E.T., Meroni, G., Merla, G. and Reymond, A. 2008. Williams-Beuren syndrome TRIM50 encodes an E3 ubiquitin ligase. *Eur. J. Hum. Genet.* 16: 1038-1049.

CHROMOSOMAL LOCATION

Genetic locus: WBSCR27 (human) mapping to 7q11.23.

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

WBSCR27 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 WBSCR27 shRNA Plasmid (h): sc-89703-SH and WBSCR27 shRNA (h) Lentiviral Particles: sc-89703-V as alternate gene silencing products.

For independent verification of WBSCR27 (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-89703A and sc-89703B.

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

WBSCR27 siRNA (h) is recommended for the inhibition of WBSCR27 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 WBSCR27 gene expression knockdown using RT-PCR Primer: WBSCR27 (h)-PR: sc-89703-PR (20 μ l). Annealing temperature for the primers should be 55-60° C and the extension temperature should be 68-72° C.