

WBSCR11 siRNA (h): sc-38621

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 WBSCR11 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. WBSCR11 is also designated GRF2IRD1, GTF3, Cream1, and MusTRD1 in human and BEN in mouse, due to slight differences in gene structure. WBSCR11 is expressed in all adult tissues as several variants and has discrete spatial and temporal expression during embryogenesis. The amino terminus of WBSCR11 interacts with transcriptional machinery proteins, while the carboxy terminus has been shown to bind the retinoblastoma protein to possibly regulate the cell cycle.

REFERENCES

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2. Pober, B.R. and Dykens, E.M. 1996. Williams syndrome: An overview of medical, cognitive, and behavioral features. *Child Adolesc. Psychiatr. Clin. N. Am.* 5: 929-943.
3. Lashkari, A., et al. 1999. Williams-Beuren syndrome: an update and review for the primary physician. *Clin. Pediatr.* 38: 189-208.
4. Osborne, L.R., et al. 1999. Identification of a putative transcription factor gene (WBSCR11) that is commonly deleted in Williams-Beuren syndrome. *Genomics* 57: 279-284.
5. Bellugi, U., et al. 1999. Bridging cognition, the brain and molecular genetics: evidence from Williams syndrome. *Trends Neurosci.* 22: 197-207.
6. Tassabehji, M., et al. 1999. A transcription factor involved in skeletal muscle gene expression is deleted in patients with Williams syndrome. *Eur. J. Hum. Genet.* 7: 737-747.
7. Bayarsaihan, D. and Ruddle, F.H. 2000. Isolation and characterization of BEN, a member of the TFII-I family of DNA-binding proteins containing distinct helix-loop-helix domains. *Proc. Natl. Acad. Sci. USA* 97: 7342-7347.
8. Yan, X., et al. 2000. Characterization and gene structure of a novel retinoblastoma-protein-associated protein similar to the transcription regulator TFII-I. *Biochem. J.* 3: 749-757.

CHROMOSOMAL LOCATION

Genetic locus: GTF2IRD1 (human) mapping to 7q11.23.

PROTOCOLS

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

PRODUCT

WBSCR11 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 WBSCR11 shRNA Plasmid (h): sc-38621-SH and WBSCR11 shRNA (h) Lentiviral Particles: sc-38621-V as alternate gene silencing products.

For independent verification of WBSCR11 (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-38621A, sc-38621B and sc-38621C.

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

WBSCR11 siRNA (h) is recommended for the inhibition of WBSCR11 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

WBSCR11 (GTF5I102): sc-81425 is recommended as a control antibody for monitoring of WBSCR11 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).

RT-PCR REAGENTS

Semi-quantitative RT-PCR may be performed to monitor WBSCR11 gene expression knockdown using RT-PCR Primer: WBSCR11 (h)-PR: sc-38621-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.