

fibrillin-1 siRNA (h): sc-43117

BACKGROUND

The fibrillin family of proteins, including fibrillin-1 (Fbn-1) and fibrillin-2 (Fbn-2), are integral components of a distinct subset of extracellular microfibrils. Microfibrils are found in elastic tissues where they facilitate elastic fiber formation and in nonelastic tissue where they support the association of the epithelial cells with the interstitial matrix. Characteristic of the fibrillin proteins are the epidermal growth factor (EGF)-like motifs which contain a consensus sequence for calcium binding. This calcium association may be critical for protein-protein interactions and stabilization of the microfibrils. Mutations of the fibrillin-1 gene have been shown to result in Marfan syndrome, a disease characterized by abnormal synthesis, secretion and matrix deposition of fibrillin. Fibrillin-2 is also linked to a rare, yet similar, skeletal disorder, congenital contractural arachnodactyly.

REFERENCES

1. Zhang, H., Apfelroth, S.D., Hu, W., Davis, E.C., Sanguineti, C., Bonadio, J., Mecham, R.P. and Ramirez, F. 1994. Structure and expression of fibrillin-2, a novel microfibrillar component preferentially located in elastic matrices. *J. Cell Biol.* 124: 855-863.
2. Zhang, H., Hu, W. and Ramirez, F. 1995. Development expression of fibrillin genes suggests heterogeneity of extracellular microfibrils. *J. Cell Biol.* 129: 1165-1176.
3. Yin, W., Smiley, E., Germiller, J., Sanguineti, C., Lawton, T., Pereira, L., Ramirez, F. and Bonadio, J. 1995. Primary structure and developmental expression of Fbn-1, the mouse fibrillin gene. *J. Biol. Chem.* 270: 1798-1806.
4. Dietz, H.C. and Peyerit, R.E. 1995. Mutations in the human gene for fibrillin-1 (FBN1) in the Marfan syndrome and related disorders. *Hum. Mol. Genet.* 4: 1799-1809.
5. Putnam, E.A., Zhang, H., Ramirez, F. and Milewicz, D.M. 1995. Fibrillin-2 (FBN2) mutations result in the Marfan-like disorder, congenital contractural arachnodactyly. *Nat. Genet.* 11: 456-458.
6. Reinhardt, D.P., Mechling, D.E., Boswell, B.A., Keene, D.R., Sakai, L.Y. and Bachinger, H.P. 1997. Calcium determines the shape of fibrillin. *J. Biol. Chem.* 272: 7368-7373.

CHROMOSOMAL LOCATION

Genetic locus: FBN1 (human) mapping to 15q21.1.

PRODUCT

fibrillin-1 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 fibrillin-1 shRNA Plasmid (h): sc-43117-SH and fibrillin-1 shRNA (h) Lentiviral Particles: sc-43117-V as alternate gene silencing products.

For independent verification of fibrillin-1 (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-43117A, sc-43117B and sc-43117C.

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

fibrillin-1 siRNA (h) is recommended for the inhibition of fibrillin-1 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 fibrillin-1 gene expression knockdown using RT-PCR Primer: fibrillin-1 (h)-PR: sc-43117-PR (20 μ l, 524 bp). 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.