

folliculin siRNA (h): sc-37412

BACKGROUND

Birt-Hogg-Dube (BHD) syndrome is a rare autosomal dominant cancer syndrome characterized by kidney tumors, benign tumors of the hair follicle and spontaneous pneumothorax. BHD is also associated with neoplastic colonic polyps. The BHD gene maps to chromosome 17p11.2 and encodes the protein folliculin. Folliculin is widely expressed. Notably, folliculin is expressed in the kidney, lung and skin, where BHD tumors arise. Specifically, the (C)8 tract in exon 11 is a mutational hot spot in BHD. BHD appears to have reduced penetrance or late onset. In a study of the renal tumors in 30 BHD patients, preoperative computed tomography scans detect a mean of 5.3 tumors per patient with a range 1-28 tumors. Multiple and bilateral tumors appear at a mean of 50.7 years.

REFERENCES

1. Balus, L., et al. 1983. Fibrofolliculoma, trichodiscoma and acrochordon. The Birt-Hogg-Dube syndrome. *Ann. Dermatol. Venerol.* 110: 601-609.
2. Schmidt, L.S., et al. 2001. Birt-Hogg-Dube syndrome, a genodermatosis associated with spontaneous pneumothorax and kidney neoplasia, maps to chromosome 17p11.2. *Am. J. Hum. Genet.* 69: 876-882.
3. Khoo, S.K., et al. 2002. Clinical and genetic studies of Birt-Hogg-Dube syndrome. *J. Med. Genet.* 39: 906-912.
4. Nickerson, M.L., et al. 2002. Mutations in a novel gene lead to kidney tumors, lung wall defects, and benign tumors of the hair follicle in patients with the Birt-Hogg-Dube syndrome. *Cancer Cell* 2:157-164.
5. Pavlovich, C.P., et al. 2002. Renal tumors in the Birt-Hogg-Dube syndrome. *Am. J. Surg. Pathol.* 26: 1542-1552.
6. Warren, M.B., et al. 2004. Expression of Birt-Hogg-Dube gene mRNA in normal and neoplastic human tissues. *Mod. Pathol.* 17: 998-1011.
7. Nagy, A., et al. 2004. Lack of mutation of the folliculin gene in sporadic chromophobe renal cell carcinoma and renal oncocytoma. *Int. J. Cancer* 109: 472-475.

CHROMOSOMAL LOCATION

Genetic locus: FLCN (human) mapping to 17p11.2.

PRODUCT

folliculin 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 folliculin shRNA Plasmid (h): sc-37412-SH and folliculin shRNA (h) Lentiviral Particles: sc-37412-V as alternate gene silencing products.

For independent verification of folliculin (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-37412A, sc-37412B and sc-37412C.

PROTOCOLS

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

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

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

folliculin (D-4): sc-271558 is recommended as a control antibody for monitoring of folliculin 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).

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgG κ BP-HRP: sc-516102 or m-IgG κ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunofluorescence: use m-IgG κ BP-FITC: sc-516140 or m-IgG κ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

RT-PCR REAGENTS

Semi-quantitative RT-PCR may be performed to monitor folliculin gene expression knockdown using RT-PCR Primer: folliculin (h)-PR: sc-37412-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.