

folliculin (C-18): sc-25168

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

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- 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.
- Khoo, S.K., et al. 2002. Clinical and genetic studies of Birt-Hogg-Dube syndrome. *J. Med. Genet.* 39: 906-912.
- 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.
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- Warren, M.B., et al. 2004. Expression of Birt-Hogg-Dube gene mRNA in normal and neoplastic human tissues. *Mod. Pathol.* 17: 998-1011.
- 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.

SOURCE

folliculin (C-18) is an affinity purified goat polyclonal antibody raised against a peptide mapping at the C-terminus of folliculin of human origin.

PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-25168 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

RESEARCH USE

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

APPLICATIONS

folliculin (C-18) is recommended for detection of folliculin of human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

folliculin (C-18) is also recommended for detection of folliculin in additional species, including equine, canine, bovine and avian.

Suitable for use as control antibody for folliculin siRNA (h): sc-37412, folliculin shRNA Plasmid (h): sc-37412-SH and folliculin shRNA (h) Lentiviral Particles: sc-37412-V.

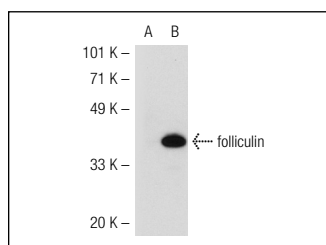
Molecular Weight of folliculin: 66 kDa.

Positive Controls: Caki-1 cell lysate: sc-2224 or folliculin (h): 293 Lysate: sc-112237.

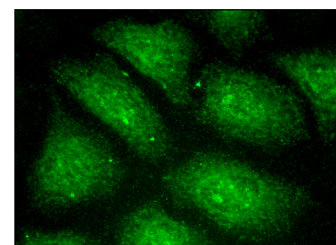
RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

DATA



folliculin (C-18): sc-25168. Western blot analysis of folliculin expression in non-transfected: sc-110760 (A) and human folliculin transfected: sc-112237 (B) 293 whole cell lysates.



folliculin (C-18): sc-25168. Immunofluorescence staining of methanol-fixed HeLa cells showing nuclear and cytoplasmic localization.

STORAGE

Store at 4° C, **DO NOT FREEZE**. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.


 MONOS
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Try **folliculin (D-4): sc-271558**, our highly recommended monoclonal alternative to folliculin (C-18).