



[Authoritative facts](#) about the skin from the [New Zealand Dermatological Society Incorporated](#).

[Home](#) | [Skin lesions](#)

Birt-Hogg-Dube syndrome

Birt-Hogg-Dube syndrome is also known as fibrofolliculoma with trichodiscoma and acrochordon. Canadian physicians Birt, Hogg and Dube first described it in 1977.

What is Birt-Hogg-Dube syndrome?

Birt-Hogg-Dube syndrome is a rare autosomal dominant inherited condition characterised by the development of benign (non-cancerous) tumours on the head, face and upper body. The three benign skin tumours involved in this condition are:

- fibrofolliculoma – tumour developing in hair follicles
- trichodiscoma – tumour of the hair disc
- acrochordon – [skin tags](#) (but these are very common and are only rarely associated with Birt-Hogg-Dube syndrome)

People with this syndrome are at increased risk for developing colon or kidney cancer as well as spontaneous pneumothorax (lung collapse) due to pulmonary (lung) cysts.

What are the signs and symptoms of Birt-Hogg-Dube syndrome?

Painless, small, papular skin lesions develop gradually over the scalp, face, neck, chest and back. Lesions usually develop at around age 30 or 40 years, although they have been found in younger patients. Features of lesions are:

- Fibrofolliculoma and trichodiscoma have similar appearance and are small (2–4mm), white-to-flesh coloured, smooth, dome-shaped bumps.
- Acrochordons or skin tags are small, soft 1–2mm bumps that look like a wart with a thin neck.
- May be anywhere from two to over a hundred lesions present.
- Once developed, skin lesions are permanent.

Oral mucosal polyps, [collagenomas](#), angioliipomas, and deforming [lipomas](#) may also form.

Birt-Hogg-Dube syndrome



What is the cause of Birt-Hogg-Dube syndrome?

Birt-Hogg-Dube syndrome is due to mutation in the gene on chromosome 17p12-q11.2 encoding a protein called folliculin. It is not known why this mutation occurs. The syndrome is an autosomal dominant condition meaning half of an affected person's children will also be affected.

How is the diagnosis made?

[Skin biopsy](#) of a lesion is performed to confirm the diagnosis. Adults with a positive biopsy result for Birt-Hogg-Dube syndrome should also undergo renal ultrasound, abdominal CT/ MRI, chest x-ray and colonoscopy to determine if there are any associated problems or malignancies.

What is the treatment for Birt-Hogg-Dube syndrome?

Patients with Birt-Hogg-Dube syndrome need to undergo medical and physical examinations and appropriate laboratory and radiographic tests on a regular basis to check for internal complications such as kidney carcinoma, pulmonary cysts, and pneumothorax.

No specific medical treatment exists for the skin lesions of Birt-Hogg-Dube syndrome. Although the skin tumours are benign they may become disfiguring if multiple lesions occur. The following skin procedures may be helpful in removing lesions, although recurrence is common.

- [Excision of skin lesions](#)
- [Dermabrasion](#)
- [Laser therapy](#)
- [Curettage and cautery](#)

Related information

References:

- [Birt-Hogg-Dube Syndrome](#) - emedicine dermatology, the online textbook

On DermNet NZ:

Other websites:

- [Birt Hogg Dube Family Alliance](#)

Books about skin diseases:

See the [DermNet NZ bookstore](#)

Author: Vanessa Ngan, staff writer

DermNet does not provide an on-line consultation service.

If you have any concerns with your skin or its treatment, see a [dermatologist](#) for advice.

Created 2005. Last updated 26 Dec 2006. © 2007 NZDS. Disclaimer.