

- Bevacizumab (Avastin) is an antibody which is used to treat bowel, lung and kidney cancer. It blocks the formation of blood vessels. Without a blood supply, cancerous cells are starved of nutrients and are not able to grow.

Other treatments are currently in development and some are undergoing clinical trials.

Considerations

The prognosis for BHD-related kidney cancer is generally positive. BHD-related cancers do not normally metastasise and so offer a good target for effective treatment. To maintain healthy kidney tissue, you may want to talk to a dietician or a nephrologist to see if they can suggest an appropriate diet. Most importantly, make sure that your kidneys are being regularly monitored by an expert clinical team.

Also available:

Birt-Hogg-Dubé Syndrome:
Clinical Introduction

Birt-Hogg-Dubé Syndrome:
Diagnosis Information

Birt-Hogg-Dubé Syndrome:
Lung Symptoms and Treatment

Birt-Hogg-Dubé Syndrome:
Skin Symptoms and Treatment

Birt-Hogg-Dubé Syndrome:
Kidney Symptoms

Birt-Hogg-Dubé Syndrome:

Kidney Treatment



BHD Birt-Hogg-Dubé Syndrome: Kidney Treatment

Introduction

Birt-Hogg-Dubé (BHD) syndrome is a rare (1 in 200,000) genetic disorder caused by alterations in the gene *Folliculin*. BHD is characterised by the development of benign skin tumours (fibrofolliculomas), lung cysts that can cause collapsed lung (spontaneous pneumothorax), and kidney cancer (renal cell carcinoma).

BHD affects people differently. If you have BHD syndrome, you may have none, one, or all of the symptoms of BHD.

Treatments

Kidney symptoms related to Birt-Hogg-Dubé syndrome might not be treated in the same manner as sporadic (non-hereditary) kidney tumours.

Most BHD-related tumours grow slowly and only need to be removed when they reach 3cm in diameter. Regular monitoring is important to keep track of the size of the tumours.

The goal in treating BHD-affected kidneys is to preserve as much kidney tissue as possible. Treatments are not permanent; it is likely new tumours will grow and may have to be removed in the future.

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Surgery

A nephrectomy is surgical removal of kidney tissue and can be full (removal of an entire kidney) or partial. Partial nephrectomies are preferable whenever possible. Nephrectomies can be laparoscopic (small incision; surgeon guided by camera) or open (full incision). Your surgeon will determine if a partial nephrectomy is a suitable treatment.



In some cases, the location or size of your tumour may make a partial nephrectomy very difficult.

Here are a few questions you may consider asking your surgeon:

- How many kidney surgeries have you performed this year? (Over 30 a year is good; over 100 a year is better)
- How many partial nephrectomies have you performed?
- If you are telling me I need a full nephrectomy, why? Can nothing be done to save part of the kidney?
- How many patients have you had with tumours similar to mine?

It is best to find an expert who is aware of the different behaviour of BHD-related tumours and who is experienced in performing the type of surgery you will need.

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Ablation

Ablation is the non-surgical removal of tissue, using heat (from radio frequency, ultrasound, or microwave) or cold (cryoablation). Ablation may be chosen as a secondary treatment if new kidney tumours grow after a nephrectomy or partial nephrectomy. Currently, ablation is not often used as a first treatment.

Biological Treatments

These treatments are used for all types of renal cell carcinoma and not specifically for BHD-related cancers.

- Interferon immunotherapy can help to stabilise or shrink a tumour by: stopping cancer cell growth, boosting the immune system to attack the cancer and restricting the blood supply to the cancer cells. Interferon- α (Roferon-A) has been used for metastatic kidney cancer.
- Interleukin 2 stimulates white blood cells (lymphocytes) to fight infection and is most often used to treat advanced kidney cancer.
- Several kinase inhibitors may be used to stop or slow the growth of kidney cancer. These include: Sunitinib (Sutent), Sorafenib (Nexavar), and Temsirolimus (Torisel).

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