

The Kidneys and BHD



Symptoms: Kidneys and BHD

Kidney tumours

BHD is associated with an increased risk of kidney cysts and tumours. Up to 1 in 3 people with BHD will get kidney cancer. It is recommended that BHD patients have regular abdominal imaging scans to detect any cancer as early as possible. Kidney tumours associated with BHD are often multifocal and can occur in both kidneys but are normally slow growing, rarely spread and not aggressive. The most common symptoms of kidney cancer include:

- Blood in your urine
- Pain in your back or side – this can vary from a sharp stabbing to persistent dull ache
- A lump or swelling in your side

Other symptoms of kidney cancer include:

- Extreme tiredness,
- Unexplained weight loss
- High blood pressure (which can also be a risk factor for kidney cancer)
- A fever and/or night sweats

Types of Kidney Tumour

The majority of BHD-associated kidney tumours are chromophobe, oncocytoma or a hybrid tumour that has areas of both of these subtypes. These types of tumours are usually slow growing and don't often spread to other areas. They are still normally removed once they reach 3 cm (1.2 inches) in size to minimize the chance of spreading (metastasizing).

Rarely, people with BHD get clear cell or papillary forms of kidney tumour. These are more aggressive and are more likely to spread. However, with regular scans, these tumours will be found and removed early, reducing the likelihood of them spreading.

Testing

To determine the type of tumour, a biopsy is taken, or the tumour is surgically removed, and is sent to a pathology lab for testing. The pathologist will also be able to determine the stage and grade of the tumour which is an assessment of how advanced the cancer is. With routine monitoring of the kidneys, the outcome for BHD-related kidney cancer is generally good.



Treatments: Kidneys and BHD

Monitoring

Most BHD-related tumours grow slowly and only need to be removed when they reach 3cm in diameter. It is strongly recommended that BHD patients have regular abdominal imaging scans to monitor their kidneys and detect any cancer as soon as possible. The type and frequency of scans may vary depending on your geographic location, healthcare service or insurance and whether or not you already have any tumours. As BHD is a genetic disease, and more tumours may develop over time, it is important to balance preservation of the kidney with early treatment of any tumours.

Treatments for BHD kidney tumours

Surgery

The most common treatment for BHD-related kidney tumours is called nephrectomy. This is a surgical method in which it is possible to remove only part of the kidney, or all of the kidney.

For BHD patients a partial nephrectomy is the preferred option as it is important to preserve as much healthy kidney tissue as possible in case additional tumours develop. There are some cases where a partial nephrectomy may not be able to remove all of the tumours and so a full nephrectomy is recommended.

Ablation

Ablation is a non-surgical method used to remove tumours and is best suited to patients with a single, small tumour. There are several types of ablation:

- Radiofrequency
- Ultrasound
- Microwave
- Cryoablation (extreme cold)

In some situations, particularly in advanced kidney cancer, additional treatments may be recommended such as tyrosine kinase (TKI) or mTOR inhibitors, and immunotherapy.

You should always inform your surgeon that you have BHD and discuss all treatment options and their side effects.

Depending on your location, the BHD Foundation may be able to recommend a surgeon experienced in treating BHD patients.

Further information

- You can read more about TKI inhibitors on the Macmillan Cancer support website
- You can read more about mTOR inhibitors on the Cancer Research UK website.
- You can read more about immunotherapy on the Cancer Research UK website

Macmillan Cancer Support website: <https://www.macmillan.org.uk/>

Cancer Research UK website: <https://www.cancerresearchuk.org/>

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If you would like to get in touch with the BHD Foundation, please email our team: contact@bhdsyndrome.org

For more information about our work and BHD, you can visit our website: www.thebhdfoundation.org