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Lung

This pamphlet tells you about the lung symptoms of Birt-Hogg-Dubé syndrome.

Birt-Hogg-Dubé Syndrome

Birt-Hogg-Dubé (BHD) Syndrome is a rare disease caused by changes in the Folliculin gene.

People with BHD can get:

1. Skin bumps (fibrofolliculomas)
2. Lung cysts
3. Collapsed lungs (pneumothorax)
4. Kidney cancer (renal cell carcinoma)

BHD affects people differently. If you have BHD, you may get none, some or all of these symptoms.

Lung cysts

- Most people with BHD get lung cysts.
- These are also called pulmonary cysts, blebs or bullae.
- Lung cysts are air-filled sacs on the outside of the lung.
- BHD lung cysts do not make breathing harder.
- Lung cysts are best seen by CT scans.

Collapsed lung

- Roughly 1 in 4 people with BHD will have a collapsed lung at least once.
- This is called a spontaneous pneumothorax.
- A small pneumothorax can sometimes heal on its own. A large or complete pneumothorax needs treatment.
- You need an X-Ray or CT scan to see if you have a pneumothorax.

The symptoms of a collapsed lung are:

1. Sharp, stabbing pain on one side of the chest
2. Pain made worse by breathing
3. Difficulty breathing
4. Dry cough

If you have any of these symptoms, **go to the hospital as soon as possible.**

Treatment - things to think about

- There are several treatments for a collapsed lung (some of which are listed on the next page).
- Each case is different. This means if you have another collapsed lung, you might get a different treatment next time.

You should talk to your doctor about the best treatment for you.

Treatment

1. Chest thoracostomy - a tube is inserted in to the chest to drain air and to let the collapsed lung reinflate by itself.
2. Pleurodesis - an operation that sticks lungs to the chest wall, stopping them collapsing again.

Which country you live in and your health insurance plan will affect which treatments you can get.

Precautions

People with BHD should talk to their doctor before flying in unpressurised planes, scuba diving, or doing certain sports.

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Produced by the BHD Foundation
Website: www.BHDSyndrome.org
Email: contact@BHDSyndrome.org



Information sources are available at:
http://www.bhdsyndrome.org/wp-content/uploads/2013/11/BHD-information-pamphlets-references_v2.pdf