



Local anaesthetic may be used to make the process more comfortable for you. A suction device may be used to speed up the draining process. Sometimes this tube will be left in place for a few days until it has been determined that the pneumothorax has resolved and the lung is functioning properly again.

Pleurodesis

A procedure that prevents fluid build up in the pleural space (the area between the membranes that surround the lungs) by attaching the outer surface of your pleural cavity to the inside of your chest cavity.

This procedure causes the membranes to stick together so there is no pleural space. This can be done chemically or mechanically. You doctor will discuss these options with you.

Pneumothorax Precautions

When you receive a positive BHD diagnosis, you may want to consult your doctor about regular surveillance of your lungs to determine whether you have lung blebs.

To find out more information,

visit :

www.BHDSyndrome.org

Also available in this series:

**BHD Syndrome:
Diagnosis Information**

**BHD Syndrome: Skin Symptoms &
Treatment**

Birt Hogg Dubé Syndrome:

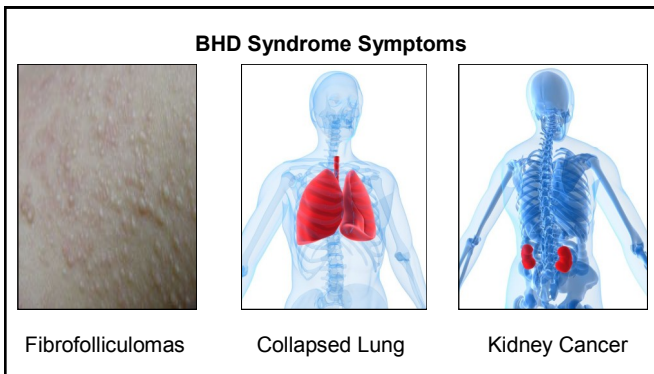
Lung Symptoms & Treatment



≡ Birt-Hogg-Dubé Syndrome: Lung symptoms & Treatment

Introduction to BHD Syndrome

Birt-Hogg-Dubé (BHD) syndrome is a rare genetic disorder characterised by the development of benign skin tumours, recurrent collapsed lung (pneumothorax) and, in some cases, kidney cancer.



There is no 'typical' BHD patient and individuals may show any, or all of these symptoms over time. BHD syndrome is caused by alterations (mutations) in the Folliculin gene.

Lung Symptoms

The lung symptoms associated with BHD Syndrome are spontaneous collapsed lung (pneumothorax), caused by the development of blebs/cysts on the surface of the lung.

A bleb is an air-filled sac on the surface of the lung, a bit like blister or an air bubble. If it ruptures, air can accumulate in the chest area causing the lung to collapse.

Lung blebs can be seen via a CT scan and many people with BHD syndrome are not aware that they have them. Even if you have blebs, you may never experience a collapsed lung.

Pneumothorax/Collapsed Lung Symptoms

Symptoms include acute chest pains, sudden shortness of breath, feeling like your heart is beating rapidly, or your skin having a slightly blue tint. If you experience any of these, you should seek a doctor's attention right away. If you cannot see a doctor immediately, go to your local Accident and Emergency department.

If you have BHD syndrome, before you have a pneumothorax, it is worth considering discussing with your doctor what to do if you think you are having a lung collapse: explain that you have Birt-Hogg-Dubé syndrome, and that once you have a pneumothorax it is likely that you will have others. This information may affect the treatment option the doctor chooses.

There is no way to predict when your lung might collapse. In the general population, tall, thin, athletic males are more likely to have pneumothoraces than other people, but this does not necessarily apply to people with BHD.

Pneumothorax Diagnosis

To determine if you are having a pneumothorax, it is likely that you will have a chest x-ray or a CT scan. Your doctor may check your lungs with a stethoscope. You may have an arterial blood gas (ABG) analysis.

Pneumothorax Treatment

Each case is different, so it is important that you discuss the best options for treatment with your doctor, and that they are aware you have BHD Syndrome.

Treatment for recurrent pneumothorax will differ with each instance.

Treatment options for collapsed lungs (pneumothoraces) will vary from case to case. Some will not need treatment. For others, your doctor may propose one of the following:

Thoracostomy

A surgical procedure that is used to release trapped air from within the chest cavity. Following a pneumothorax, an incision is made in your chest so a tube can be inserted to drain air from this area.

Once the trapped air is removed, the lung can often re-expand on its own. The doctor will determine if this is an option for your pneumothorax.