

### Thoracotomy and VATS

A thoracotomy is a surgical procedure that opens the chest cavity by making an incision through the chest wall. This is used to look more closely at the lungs, and to remove a lung or part of a lung (lung resection). VATS (video-assisted thoracic surgery) is less invasive, as surgeons make a smaller incision and use cameras to look inside the chest cavity. Both thoracotomy and VATS are used to perform pleurodesis.

### Precautions

Flying in unpressurised aeroplanes, scuba diving, certain sports, strenuous activity at high altitudes, and smoking may increase the risk of spontaneous pneumothorax. However, the increase in risk is different for each individual and it may not be necessary to avoid such activities.



We recommend talking to your doctor about your BHD symptoms. You can discuss how you can avoid a pneumothorax, and what you should do if you think you are having a pneumothorax, so you are prepared to act appropriately.

### Also available:

Birt-Hogg-Dubé Syndrome:  
Clinical Introduction

Birt-Hogg-Dubé Syndrome:  
Diagnosis Information

Birt-Hogg-Dubé Syndrome:  
Skin Symptoms and Treatment Options

Birt-Hogg-Dubé Syndrome:  
Kidney Symptoms

Birt-Hogg-Dubé Syndrome:  
Kidney Treatment Options

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Produced by the BHD Foundation.  
Email: [contact@BHDSyndrome.org](mailto:contact@BHDSyndrome.org)

## Birt-Hogg-Dubé Syndrome:

## Lung Symptoms and Treatment Options



# BHD Birt-Hogg-Dubé Syndrome: Lung Symptoms and Treatment Options

## Introduction

*This pamphlet offers an overview of the lung symptoms linked with Birt-Hogg-Dubé syndrome (BHD) and the treatment options available.*

BHD 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.

## Lung Symptoms

The lung symptoms associated with BHD are cysts and spontaneous pneumothorax.

The cysts (also called lung blebs or bullae) are sacs in the lung tissue that are filled with air. Cysts can be detected by CT scans, though regular monitoring is not required. You may not be aware that you have lung cysts before having a scan.

The cysts may rupture. If that happens, the air from the ruptured cyst enters the space in between the lung and the chest wall (called the pleural space), creating pressure on the lungs which can lead to a spontaneous pneumothorax.

## Spontaneous Pneumothorax

Spontaneous pneumothorax may be partial or complete. A very small pneumothorax may heal on its own, but it is important to see a doctor if you think you are having a pneumothorax. A complete pneumothorax will require medical attention.

Symptoms may include: chest pains and shortness of breath; sensation of constricted chest; urge to cough (dry cough); rapid heartbeat.



The symptoms of a pneumothorax may feel similar to a heart attack or other conditions.

Only a doctor can determine whether or not you are having a pneumothorax.

If you experience these symptoms, you should go to an Urgent Care/ Emergency Department in a hospital. A doctor will usually perform a chest X-ray, CT scan or ultrasound to see if you are having a pneumothorax. Your doctor may also check your lungs with a stethoscope and you may have an arterial blood gas analysis.

## Pneumothorax Treatment

Each pneumothorax case is different, so it is important that you have expert help to determine what treatment is best for you. If you have had more than one pneumothorax, the treatment will depend on each instance and may not be the same. A smaller pneumothorax may only require a small needle aspiration to drain air, rather than the procedures below. Additionally, any treatment carries a risk of side effects. You may wish to discuss potential effects with your doctor.

### Chest thoracostomy

A thoracostomy is a surgical procedure in which a tube is inserted to drain air from the chest cavity. The lung can then often re-expand on its own.

During a chest thoracostomy, a local anaesthetic may be used to make the procedure more comfortable

### Pleurodesis

This procedure causes the membranes that surround the lungs to stick to the lungs themselves, eliminating the pleural space and preventing air accumulation. There are several ways a pleurodesis can be performed, for example by inserting chemicals such as talc into the pleural space. Pleurodesis is combined with thoracostomy, thoracotomy or VATS; there is evidence that it may be more effective if combined with either thoracotomy or VATS.