PULMONARY HYPERTENSION CLINIC

Fortis

FREQUENTLY ASKED QUESTIONS

1 WHAT DOES PULMONARY HYPERTENSION MEAN?

Pulmonary hypertension is a form of **Hypertension ("High Blood Pressure")** in the vessels of the body carrying blood from the lungs to the heart. It is different from routine "high blood pressure" in the fact that, hypertension refers to the higher pressures in vessels delivering blood from the heart to the rest of the body.

2 HOW DOES PULMONARY HYPERTENSION DEVELOP?

Pulmonary hypertension can occur due to various reasons in a individual including a change in the 'make' of the vessels (their walls), a back pressure from the heart, a physical occlusion in the vessels or at times due to unknown reasons. Imagine this to be a set of pipes delivering water to a housing unit to understand this easily, except think of these as rubber pipes. So, if there are several pipes coming to the house, if there is a block at the faucet in the house, the pressure will be felt in each of these pipes back. If some of the rubber pipes are smaller than the others, their ability to dilate and sustain this pressure will obviously be different.

Similarly, if some rubber pipes are stiffer than the rest it will make the pressures unequal. Additionally, if there is silt or dirt collected in some pipe the pressure will be affected. The pipes are the pulmonary vessels, the rubber make and the stiffness is the differing elasticity of the vessels and the silt is the occlusion.

3 WHICH MEDICAL CONDITIONS ARE ASSOCIATED WITH OR CAUSATIVE FOR PULMONARY HYPERTENSION?

Conditions causing or associated with Pulmonary hypertension are classified into **5 groups** internationally:

Group 1: Pulmonary Arterial Hypertension

In this group, the pipes (vessel walls) are stiffer than usual due to a variety of causes

- Idiopathic – unknown cause
- Hereditary – due to inherited genes from your family
- PAH with other medical conditions – due to diseases like HIV, liver cirrhosis, some connective tissue diseases, schistosomiasis, congenital heart disease
- Medications – some medications including some for cancer treatment can be causative

Group 2: Pulmonary hypertension due to left heart disease

In this group, like in the earlier analogy, the faucet is the problem and thereby there is back-pressure on the pipes. The left compartment of the heart receives blood from the lungs and consequently inability to effectively push blood further can cause such "back-pressure". This can be due to weakening or stiffening of the heart musculature due to coronary artery disease (blockages in the blood vessels of the heart), due to valvular disorders (certain key gateways in the passage of blood within the heart malfunction), other conditions where the heart muscle is stiffer due to an internal cause like a connective tissue disease (systemic sclerosis for example) or simply overloading the heart . Most patients belong to this group.

Group 3: Pulmonary hypertension due to lung disease and/or hypoxia

Several lung conditions can cause some remodelling in the vessels of the patient due to chronically lower oxygen or structurally. Common conditions include chronic obstructive pulmonary disease and interstitial lung disease. Similarly in disorders like obstructive sleep apnea (simplistically a disorder with loud snoring or nocturnal choking events) the oxygen level fluctuations may be akin a chronic lung disease and can in various ways contribute to the development of pulmonary hypertension.

Group 4: Pulmonary hypertension associated with pulmonary artery obstructions

In these patients, blood clots form and persist due to various reasons in the pulmonary vessels. The 'blockages' in the 'pipes' consequently lead to raised pressures. Other causes could be cancers which cause blockages within the vessels and some other rare causes for physical obstructions within the vessels.

Group 5: Pulmonary hypertension with unclear or multifactorial mechanisms

This group is a miscellaneous group of patients in whom we may not always know the mechanism of development of higher pressures or we cannot attribute a single mechanism. This includes a variety of diseases including blood disorders like sickle cell anaemia, chronic kidney disease, sarcoidosis, fibrosing mediastinitis and more.

4 WHAT ARE THE SYMPTOMS OF PULMONARY HYPERTENSION?

Symptoms of pulmonary hypertension can appear gradually over time as the pressures increase. Some may show sudden symptoms because of some other condition worsening hitherto undiagnosed pulmonary hypertension. ***These symptoms include:***

- Shortness of breath
- Syncopal episodes (fainting) or pre-syncope (light headedness)
- Breathlessness on bending forward
- Chest pain
- Swelling over your feet and abdomen
- Bluish discoloration of lips and finger tips
- Palpitations (self-awareness of a quick or pounding heart beat)
- Coughing out blood

While these symptoms may be seen in other ailments as well, it is essential to notify your care provider when you notice them so appropriate investigations may be done to rule out pulmonary hypertension.

5 HOW IS PULMONARY HYPERTENSION DIAGNOSED?

You may be required to undergo a variety of tests to identify what has caused the pulmonary hypertension, which may include the following:

- Electrocardiogram
- 2D ECHO screening
- CT scans of the chest
- Pulmonary function tests (where you are needed to blow forcefully into a machine)
- A sleep study (to check for 'snoring disorders')
- Right heart catheterization (a process where a they check the pressures in the pulmonary vessels)
- Arterial blood gas (checking blood oxygen levels)

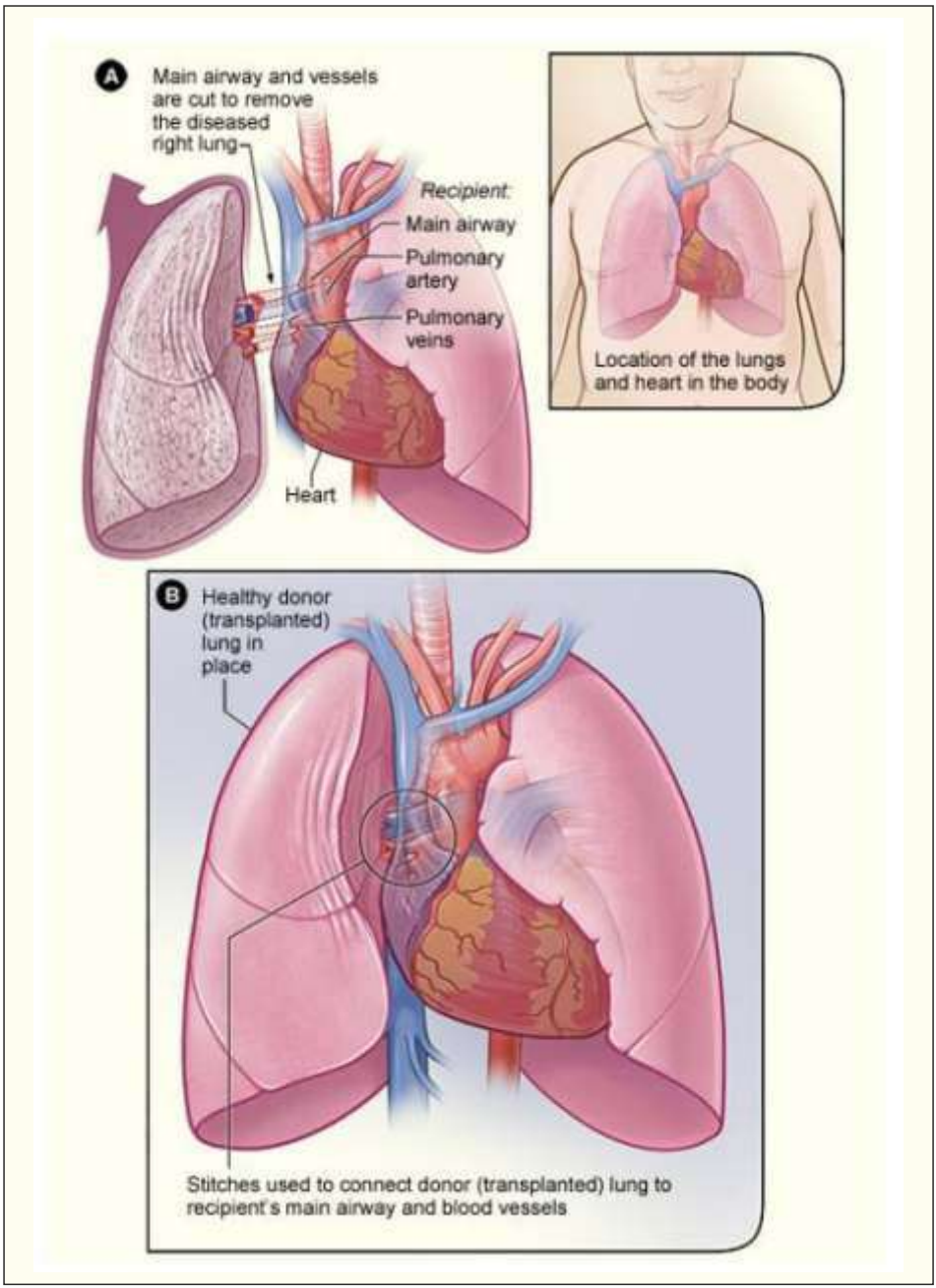
- Blood tests to assess the stress on the heart or understand underlying diseases

While this is not an exhaustive list, your physician will likely ask for several tests and may need to refer you to other physicians of different faculties like rheumatology, haematology and cardiovascular thoracic surgery, which may help narrow down the cause of the pulmonary hypertension and some to further identify the severity of the disease as well.

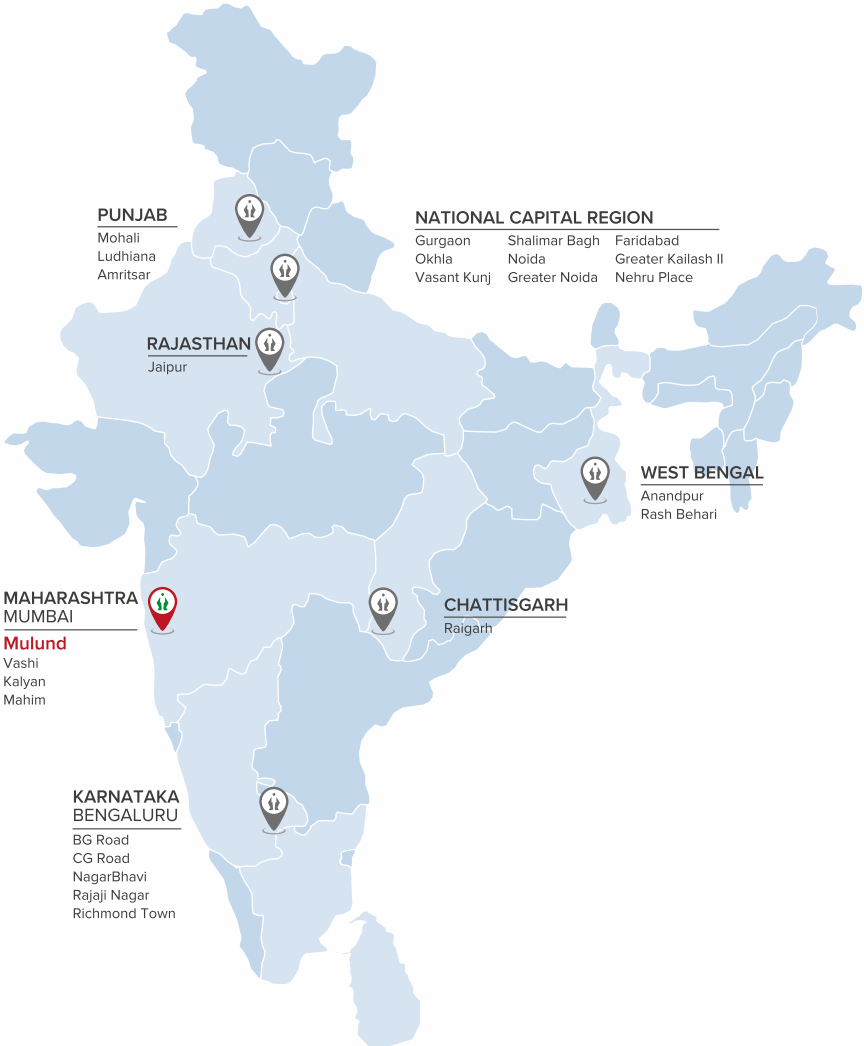
6 HOW IS PULMONARY HYPERTENSION TREATED?

If you suffer from pulmonary hypertension, it is vital to know that there are different therapies based on the different causes of pulmonary hypertension and you must visit a physician or centre with appropriate experience with dealing with the same. This may include medical management for the underlying diseases, medicines to reduce the pressures in the vessels themselves, blood thinning medicines, surgical techniques for improving right ventricular function or even transplanting a new heart and lungs in extreme cases.
There is significant role of limited daily physical exercises. Yoga and breathing exercises also help in improving lung capacity and reduction of symptoms.

Visit a pulmonary hypertension clinic to understand your choices and what is most appropriate for you


A
Main airway and vessels are cut to remove the diseased right lung
Recipient:
Main airway
Pulmonary artery
Pulmonary veins
Heart
Location of the lungs and heart in the body
B
Healthy donor (transplanted) lung in place
Stitches used to connect donor (transplanted) lung to recipient's main airway and blood vessels

THE FORTIS HOSPITAL NETWORK



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