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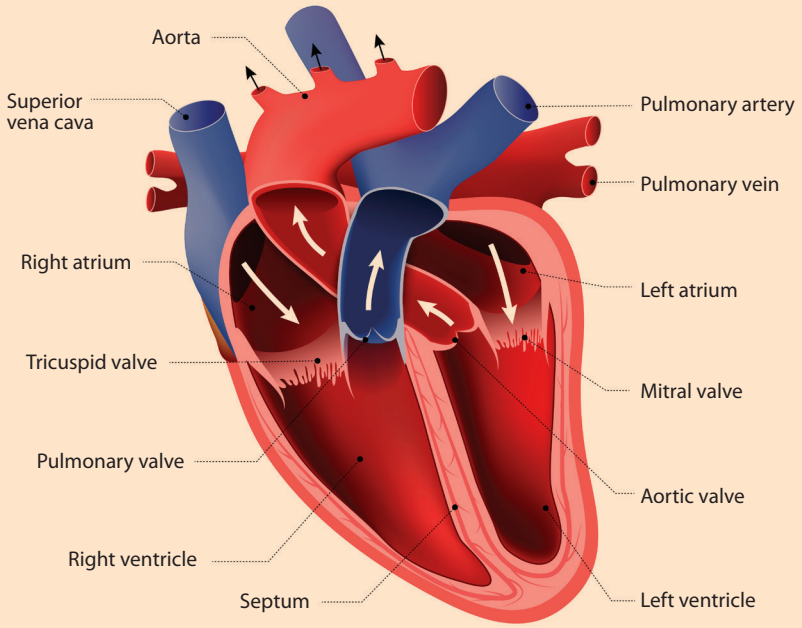
Pulmonary arterial hypertension

Changi General Hospital
SingHealth

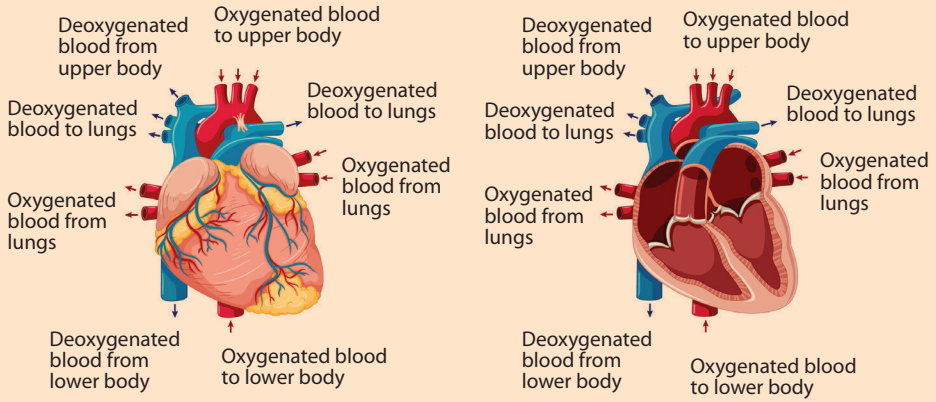
What is pulmonary arterial hypertension (PAH)?



Pulmonary arterial hypertension (PAH) occurs when the blood vessels in the lungs are diseased and become thick and narrow, resulting in an increase of pressure on the right side of the heart as it tries to pump blood through these narrow blood vessels.

To understand pulmonary arterial hypertension (PAH), you need to know how blood circulates throughout your body.

The heart works like two pumps connected to each other. The left and right side of the heart each have different roles.



The left side of the heart (left atrium) receives oxygen-rich blood from the lungs, while the left ventricle pumps this blood throughout your body. As the left side of the heart has to pump large volumes of blood over a great distance, it is designed to pump against relatively high pressure. This pressure is measured easily with a blood pressure cuff and is called your **systemic blood pressure** or **blood pressure**. When your systemic blood pressure is too high, it is called **systemic hypertension** or simply, **hypertension**.

After your blood has delivered oxygen to the organ tissues of your body, the blood becomes oxygen-poor and is then circulated to the lungs for more oxygen. This happens by sending blood from the rest of the body to the right side of the heart (right atrium), so the right ventricle pumps blood into the lungs, and oxygen enters the bloodstream. The blood does not need to travel far to move from the right side of the heart to the lungs, so the right side of the heart pumps against relatively lesser pressure. The pressure that the right side of the heart pumps against is usually a low-pressure system called **pulmonary pressure**. When this pressure gets too high, **pulmonary hypertension** (PH) occurs.

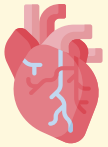
Are pulmonary hypertension and pulmonary arterial hypertension the same condition?

No, pulmonary hypertension (PH) and pulmonary arterial hypertension (PAH) are not the same. PAH is a subtype of PH.

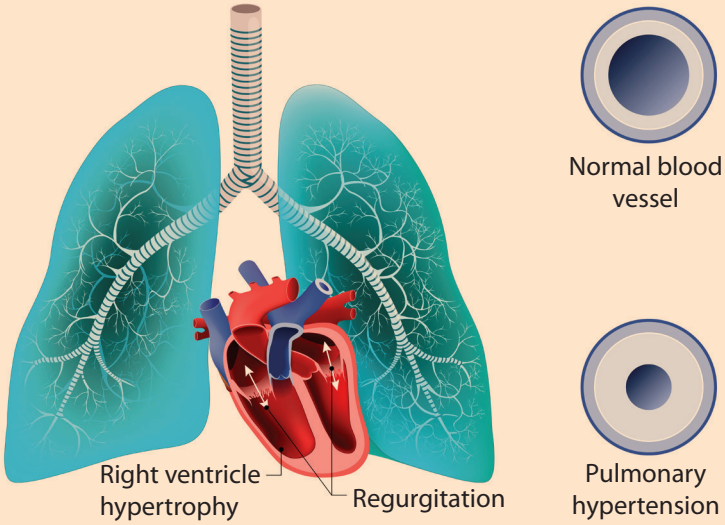
Pulmonary hypertension (PH) is a general term used to describe elevated blood pressure on the right side of the heart, but it does not explain the cause for this elevation. This high pressure could be caused by various medical conditions such as:

Chronic lung disease

Blood clots in the blood vessels in the lungs (chronic thromboembolism)



Left-sided heart problems (causing a backup of pressure on the right side of the heart)



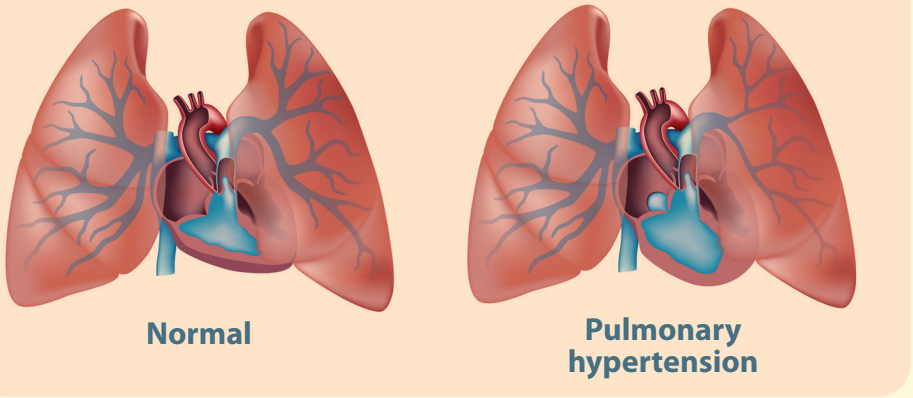
In PAH, the increase in pressure from the right side of the heart is usually much higher than the pressure in patients with PH from other causes. This results in significant breathlessness and even right heart failure.

What happens to the body with pulmonary arterial hypertension?

With PAH, the arteries become too narrow to handle the amount of blood that needs to be pumped through the lungs.

As a result, there will be:

- A backup of blood in the veins returning blood to the heart
- An increase in the pressure that the right side of the heart has to pump against
- A strain on the right side of the heart due to the increased work that it has to do to push blood through the lungs



If PAH is left untreated, there will be inflammation and remodelling of the blood vessels, and the pulmonary arteries (blood vessels of the lung) become increasingly narrow, thicker and less flexible. This narrowing of the pulmonary arteries increases the pulmonary pressure, causing the right side of the heart to become overworked, and may eventually lead to failure. The blood oxygen level may be lower than normal, as the blood has difficulty getting through the lungs to pick up oxygen. This low oxygen level strains the heart and decreases the amount of oxygen delivered to the brain and other organs.

What are the symptoms of pulmonary arterial hypertension?

The symptoms of PAH may share similarities with symptoms of other illnesses. This can make it difficult for doctors to identify PAH.

Common symptoms include:


Shortness of breath during physical or normal activities


Extreme tiredness (fatigue)


Fast heartbeat (palpitations)


"Light-headedness" or dizziness


Fainting (syncope)


Chest pain or discomfort


Dry cough


Swelling in the legs (oedema)


Swelling of the belly (ascites)

PAH can affect anyone of any age, sex or race. It is, however, more common in women between 30 and 50 years old.

How is pulmonary arterial hypertension diagnosed?

As treatment of the different pulmonary hypertension subtypes vary, patients should inform their doctor of the type and progression of their symptoms (including, but not limited to those listed in the previous section).

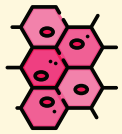
If the doctor suspects that the patient has PAH, they will order tests to determine if there is a strain on the right side of the heart. The initial tests are:

- Blood test, called NT-pro-BNP
- A chest x-ray
- Computed tomography (CT) of the lungs
- Electrocardiogram (ECG)
- Lung function tests
- An ultrasound scan of the heart (echocardiogram)
- If the test results suggest high pressure on the right side of the heart, the doctor may order a cardiac catheterisation (on the right and possibly left side of the heart as well)

The use of a right heart catheterisation is the best way to measure the blood pressure in the right side of the heart. It also provides additional information on the specific subtype of pulmonary hypertension (refer to the CGH brochure on Cardiac Catheterisation for more details).

What causes pulmonary arterial hypertension?

PAH can be inherited (*familial*), or caused by reasons that are unknown (called *idiopathic* PAH). Other known causes of PAH (called *associated* PAH) include:



Connective tissue diseases (autoimmune conditions) e.g. systemic sclerosis (also known as scleroderma), systemic lupus erythematosus (SLE), or mixed connective tissue disease (MCTD)

Prescription medications such as diet pills

Improper drug use (cocaine, methamphetamines)

Congenital heart defects

Liver disease/cirrhosis

Human Immunodeficiency Virus (HIV)

The cause of PAH is difficult to determine, and doctors will likely order several tests to find the cause. These may include:

- Blood tests including (but not limited to) autoimmune serologies, HIV screening, drug toxicologies, hepatitis serologies
- Nailfold capillaroscopy (refer to the CGH patient brochure on Nailfold Capillaroscopy)
- If an associated condition is suspected, your heart specialist may arrange for a referral to other specialist doctors such as the rheumatologist (autoimmune doctor), respiratory physician, gastroenterologist, and/or an infectious disease specialist for further checks

Is there a cure for pulmonary arterial hypertension?

PAH is a progressive and serious disease that has no cure at this time. However, there are treatments available to manage the condition.

It is important that both patient and doctor discuss the condition and treatment options together, as the disease and response to therapy differs for each patient.

Early diagnosis and treatment are essential in limiting the progression of PAH.

How is pulmonary arterial hypertension treated?

Treatment options for patients with PAH can be broadly classified into general measures, supportive measures and PAH-specific therapies.

General measures

These include lifestyle modifications such as the avoidance of strenuous activities, careful family planning and the use of appropriate contraception, and vaccinations for influenza and pneumococcus.

Supportive measures

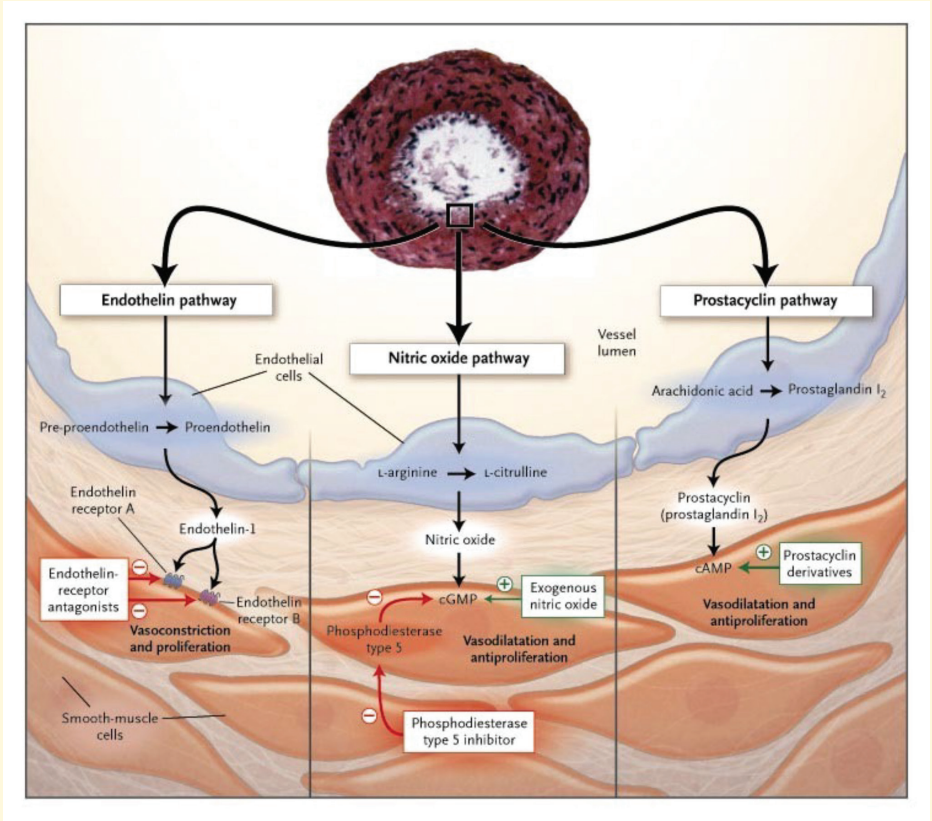
These include both pharmacological and non-pharmacological means.

- Pharmacological measures e.g. oral anticoagulants help to prevent thromboembolism (blood clot formations), while diuretics or "water pills" help to remove excess fluid that tends to build up in the body and cause organ congestion.
- Non-pharmacological measures such as oxygen supplementation and pulmonary rehabilitation improve breathing and patient activity levels.

PAH-specific therapy

Medications used to treat PAH help open the blood vessels in the lungs, thus improving blood flow through the lungs and reducing the strain on the heart. There are several types of treatment available for PAH, which target various pathways:

- **The Nitric Oxide Pathway** - includes drugs such as sildenafil, tadalafil and riociguat.
- **The Endothelin Receptor Pathway** - includes drugs such as bosentan, macitentan, and ambrisentan.
- **The Prostacyclin Pathway** - includes drugs such as selexipag, inhaled iloprost, iv prostacyclin etc.



Source: Humbert M et al. N Enal J Med 2004: 351:1425-1436

Current management and treatment guidelines for pulmonary arterial hypertension advocate the early introduction of combination therapy using drugs that act on two or even all three of the possible pathways. Such treatment strategy has been shown to improve the patient's symptoms and risk profile, as well as long-term clinical outcome.

New medications are being studied and may be suggested or recommended for the treatment of PAH as part of clinical research trials. Please consult with your doctor for more information.