

# Group I Pulmonary Hypertension

## What is Group I Pulmonary Hypertension (Pulmonary arterial hypertension)?

**Pulmonary arterial hypertension**, or Group I pulmonary hypertension (PAH or Group I PH) is a type of pulmonary hypertension (high blood pressure in the lungs) that is specifically due to narrowing of the pulmonary arteries (blood vessels) that carry blood to the lungs.

Pulmonary arterial hypertension is a broad category which includes many patients with different types of PAH. These include:

- Idiopathic
- Heritable (genetic)
- Associated with connective tissue disease
- Associated with other conditions

## What causes Group I pulmonary hypertension?

The exact cause of pulmonary arterial hypertension is not well understood. Narrowing of pulmonary arteries in PAH is due to multiplication and growth of cells in the walls of the pulmonary arteries, as well as scar tissue formation in the pulmonary arteries. Tremendous ongoing research has identified many biologic abnormalities in pulmonary arterial hypertension, including genes, proteins, and small chemicals in the body.

In many cases, pulmonary arterial hypertension is associated with another medical condition.

Some patients inherit pulmonary hypertension from their parents. Patients with inherited pulmonary arterial hypertension are said to have **heritable pulmonary arterial hypertension** (HPAH). HPAH should be suspected when more than one closely related person in a family (parent, child, sibling) has pulmonary arterial hypertension. Even in a family with known HPAH, the risk to individuals is still quite small: on average, only 1 in 10 will develop pulmonary arterial hypertension.

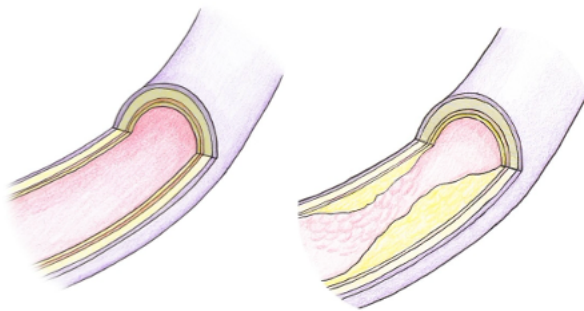
It is important to realize that in most patients with pulmonary hypertension (all types other than HPAH), there is no evidence that pulmonary hypertension is inherited.



# What happens when you have Group I pulmonary hypertension?

For our bodies to function optimally, we need oxygen to be delivered to each and every organ. Our lungs provide oxygen to the red blood cells, which then deliver it to the rest of the tissues and organs. To achieve this, blood flows through a network of vessels which distribute the blood throughout the lungs. These vessels are lined on the inside with cells known as **endothelial cells**. In the blood vessels of a person with healthy lungs, the endothelial cells follow a strict life cycle: they form, multiply, die and are replaced.

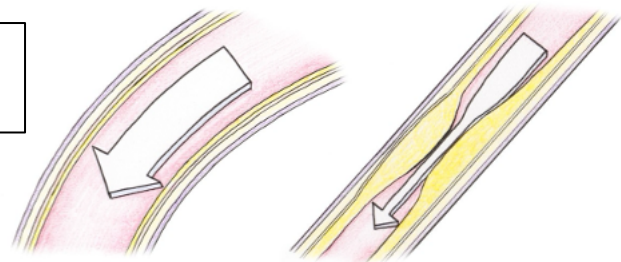
In pulmonary arterial hypertension (PAH), this life cycle is disrupted. The endothelial cells multiply extensively without dying off. This causes them to build up in layers on the walls of the blood vessels. This gradually makes the blood vessels narrower. Smaller vessels can become completely clogged. The excess endothelial cell buildup also makes the walls of the blood vessels stiffer and less flexible.



Healthy blood vessel (left) vs. PAH-affected blood vessel (right)

When the blood vessels in the lungs become narrower and stiffer, there is less room for blood to flow through.

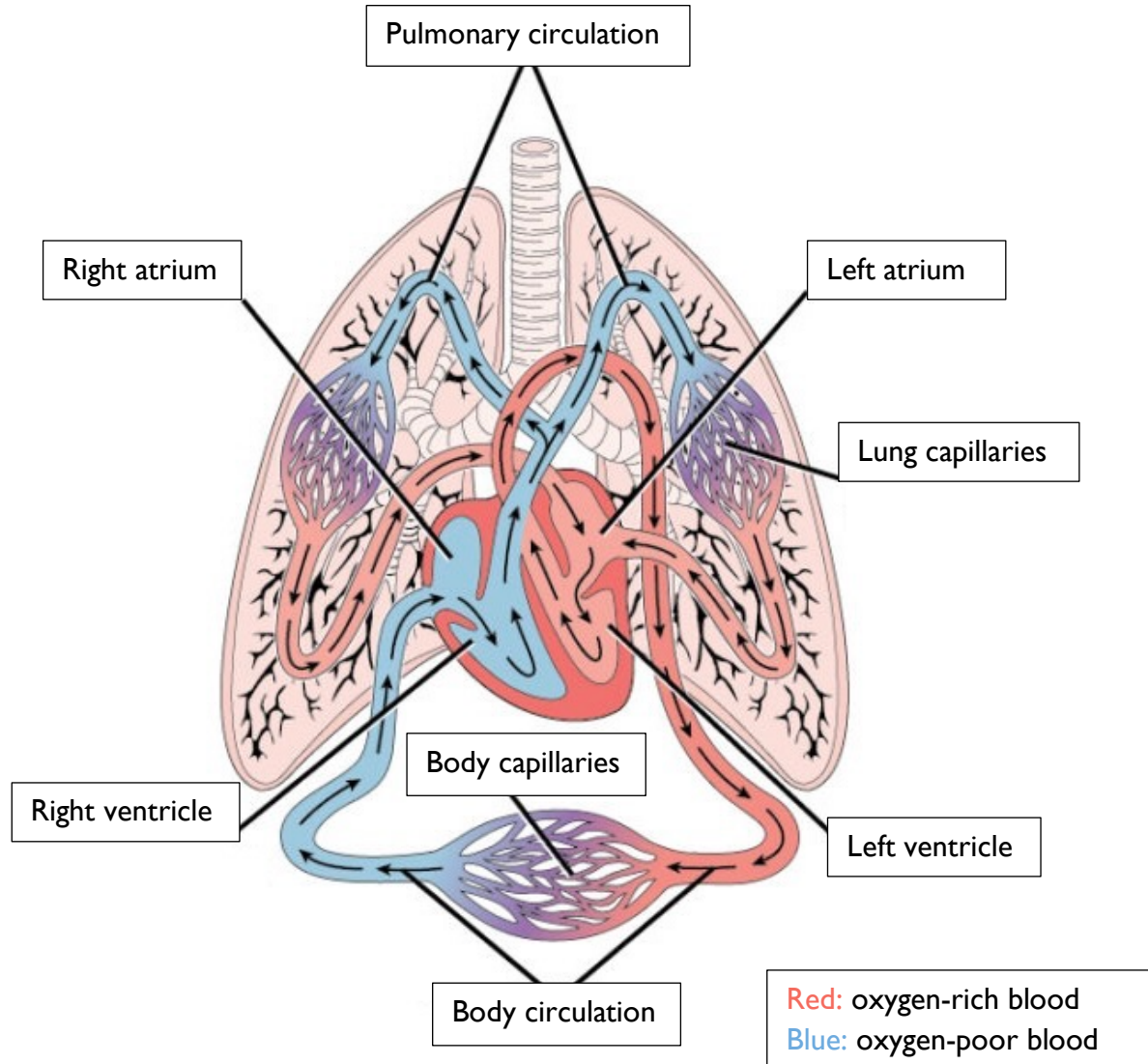
Normal blood flow (left) vs. reduced blood flow in narrowed blood vessels (right)



This reduced blood flow weakens the lungs' ability to oxygenate the blood, which can result in lower blood oxygen levels throughout the body.



The right side of the heart pumps blood through the lungs.



Because of the narrowing of the blood vessels in the lungs, the heart must work harder to push blood throughout the lungs. Imagine trying to pump water through a wide, clean pipe versus a clogged pipe. The same happens in the lungs when the blood vessels are narrowed.

As the right heart works harder, over time it becomes enlarged and weakened. This places people living with pulmonary arterial hypertension (PAH) at risk of right heart failure, and leads to symptoms including swelling, nausea, chest heaviness and/or palpitations.



## Diagnosis

Echocardiography, pulmonary function tests, and arterial blood gases play a role, but a procedure called a **right heart catheterization** is the only way to definitively diagnose pulmonary arterial hypertension.

During a right heart catheterization, the doctor will calculate how much blood the heart can pump in a minute (cardiac output) and take a reading called a “wedge” pressure. In this test, a balloon is inflated at the end of the catheter and wedged into a smaller section of one of the pulmonary arteries. Patients with pulmonary arterial hypertension (Group I PH) will have normal wedge pressure.

Elevation in the mean pulmonary artery pressure (mPAP) confirms pulmonary hypertension and can also exclude dysfunction of the left heart as a contributor to the pulmonary hypertension.

## Treatment

Since 1997, 11 pulmonary hypertension-specific medical treatments have been approved in Canada for the treatment of pulmonary arterial hypertension (PAH). Your pulmonary hypertension specialist will work with you to determine which treatment options best meet your needs.

### Endothelin receptor antagonists

Patients with pulmonary arterial hypertension make excess endothelin, one of the most powerful chemicals which constricts blood vessels. This causes cells to multiply, narrowing pulmonary arteries and increasing pulmonary arterial pressure. Endothelin receptor antagonists (ERAs) block the action of this excess endothelin opening up pulmonary arteries and reducing pulmonary arterial pressure, as well as improving right ventricular function. ERAs also make cells stop multiplying (they are anti-proliferative) and decrease scar tissue formation (they are anti-fibrotic).

Endothelin receptor antagonists include ambrisentan (Volibris®), bosentan (Tracleer®), and macitentan (Opsumit®).

### Phosphodiesterase inhibitors and soluble guanylate cyclase stimulators

Patients with pulmonary arterial hypertension make less nitric oxide (NO), which is a normal pulmonary vasodilator. Phosphodiesterase inhibitors (PDE-5 inhibitors) and soluble guanylate cyclase stimulators (sGC stimulators) are medications which improve the action of patients' own nitric oxide. They are pulmonary vasodilators that can open up pulmonary arteries and reduce pulmonary arterial pressure, as well as improve right-side heart function.



Phosphodiesterase inhibitors include sildenafil (Revatio®) and tadalafil (AdCirca®).

Soluble guanylate cyclase stimulators include riociguat (Adempas®).

### IP receptor agonists and prostacyclin analogues

Patients with pulmonary arterial hypertension make less prostacyclin, which is a normal pulmonary vasodilator. Chemicals like prostacyclin (IP receptor agonists and prostacyclin analogues) are also pulmonary vasodilators that can open up pulmonary arteries and reduce pulmonary arterial pressure, as well as improve right-side heart function. They also make cells stop multiplying (they are anti-proliferative) and decrease blood clot formation (they are anti-platelet).

IP receptor agonists include selexipag (Uptravi®).

Prostacyclin analogues include epoprostenol (Flolan®, Caripul®) and treprostinil (Remodulin®).

### Activin signalling inhibitors

The pathway associated with BMPR2 signalling is important in controlling cell growth in pulmonary arteries. In patients with pulmonary arterial hypertension, this signal seems to be under-active, and a competing pathway known as the activin pathway becomes predominant and stimulates abnormal cell growth. The activin signalling inhibitor binds the activins and inhibits abnormal cell proliferation in the pulmonary arteries.

Activin signalling inhibitors include sotatercept (Winrevair®).

## Other treatments

### Oxygen therapy

Patients may require supplemental oxygen to maintain adequate oxygen levels. Typically, the target is an oxygen saturation above 90%, but this will vary depending on the condition.

### Cardiopulmonary rehabilitation

Cardiopulmonary rehabilitation can help symptoms and increase exercise capacity for people with many types of pulmonary hypertension.

### Transplant

Depending on the severity of pulmonary hypertension, heart or lung transplantation might also be an option.



## Key points for patients

- Learn about the management of your condition from your pulmonary hypertension specialist.
- Obtain a pulse oximeter. Learn how to monitor oxygen levels and how to adjust your supplemental oxygen (if applicable).
- Learn about what exercise you can safely do and do it to keep your muscles active.
- Stay up to date on all your medications and vaccinations.
- Avoid triggers that exacerbate pulmonary hypertension, including anemia, infection, high salt intake, and straining.
- Watch for fluid retention and report it to your care team.
- Ask your care team about any clinical trials in Group I PH. Ongoing investigations are being conducted to improve outcomes in this condition.

## Image credits

Pulmonary circulation image:

Rye C, Wise R, Jurukovski V, DeSaix J, Choi J, & Avissar Y. (2016). *Biology*. OpenStax. [https://openstax.org/books/biology/pages/40-1-overview-of-the-circulatory-system#fig-ch40\\_01\\_03](https://openstax.org/books/biology/pages/40-1-overview-of-the-circulatory-system#fig-ch40_01_03) used under a Creative Commons Attribution license

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