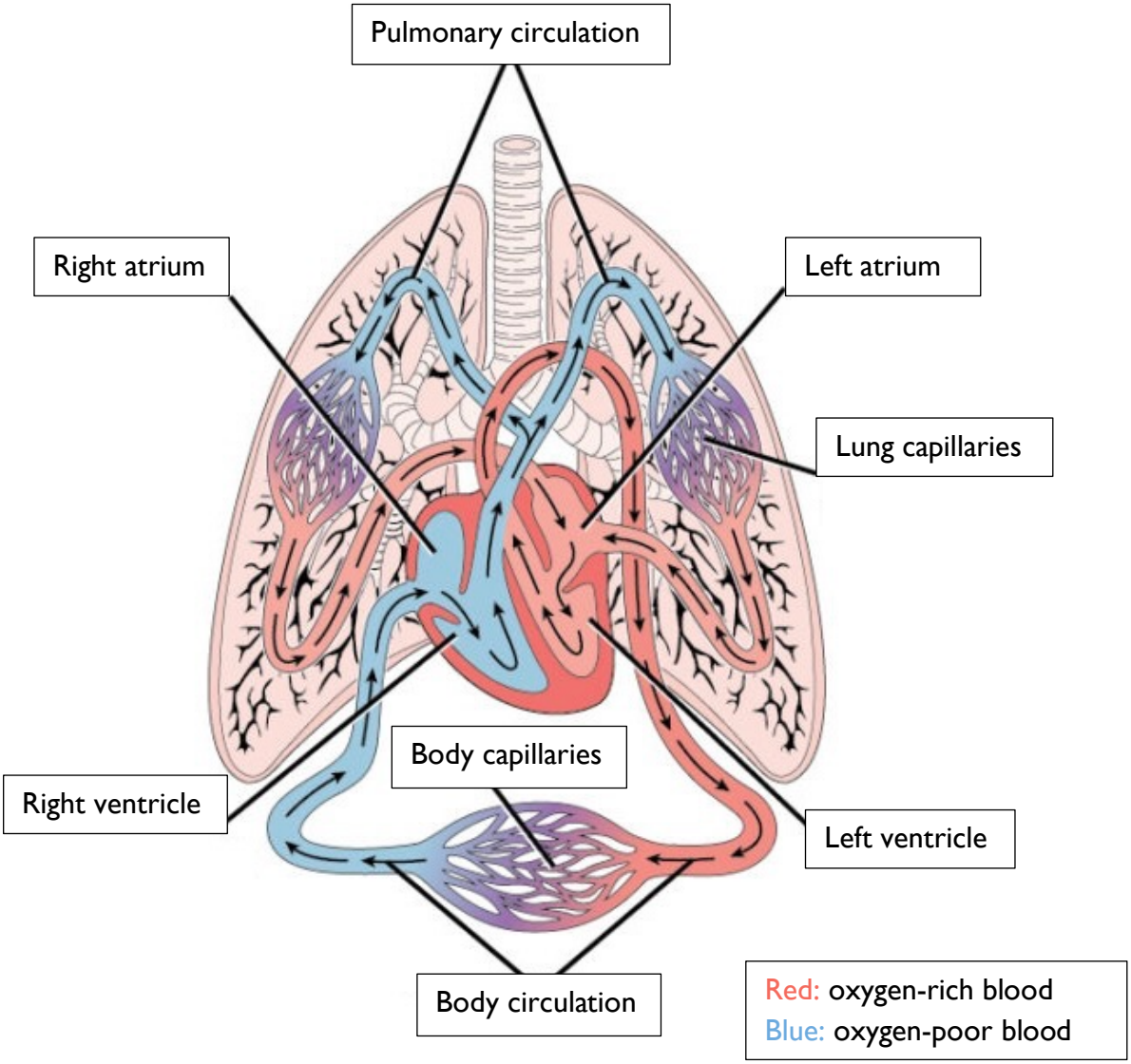
Group 5 Pulmonary Hypertension

What is Group 5 pulmonary hypertension?

Group 5 pulmonary hypertension (Group 5 PH) is pulmonary hypertension (high blood pressure in the lungs) that arises from various other diseases.

What happens when you have PH?

High blood pressure throughout the lungs puts a progressive strain on the right side of the heart.



PULMONARY HYPERTENSION ASSOCIATION OF CANADA
L'ASSOCIATION D'HYPERTENSION PULMONAIRE DU CANADA

www.phacanada.ca
info@phacanada.ca | 1-877-774-2226
A better life for all Canadians affected by pulmonary hypertension

Last reviewed: 2026

What causes Group 5 pulmonary hypertension?

The most common causes include:

Blood-related disorders

Some blood-related disorders can cause pulmonary hypertension, including:

- Polycythemia vera (when your bone marrow makes too many red blood cells)
- Essential thrombocythemia (when your bone marrow makes too many platelets)
- Sickle cell anemia (when red blood cells are sticky and misshapen)
- Chronic hemolytic anemia, also called thalassemia (when blood cells die off faster than they can be replaced)
- Splenectomy (spleen removal).

Systemic disorders

Systemic disorders that involve the lungs can cause pulmonary hypertension, including:

- Sarcoidosis, (a complex immune disease that can result in inflammation, scarring, and damage to tissues in the lungs and blood vessels)
- Lymphangioleiomyomatosis (when too much smooth muscle growth causes cysts in the lungs)

Kidney disease

A significant proportion of people with chronic kidney disease develop pulmonary hypertension.

Certain metabolic disorders

Examples include:

- Glycogen storage disease (when the body has difficulty breaking down certain sugars)
- Thyroid disease
- Gaucher disease (a genetic disorder in which fat cells build up in certain organs)

Cancers

Some cancers can cause pulmonary hypertension, if tumours press on the pulmonary arteries.

Other rare diseases

Other rare diseases, including mediastinal fibrosis (a disease resulting in progressive scarring of the tissues in the area between the lungs and the chest wall), can cause pulmonary hypertension.

Diagnosis

The diagnosis relies on a clinical probability assessment. Echocardiography, pulmonary function tests, and arterial blood gases play a major role.

Some patients require a procedure called a **right heart catheterization**. 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

Treat the underlying disease

The primary goal of treatment is to manage the underlying disease and address any hypoxia (low oxygen level in the blood), which can help improve the pulmonary hypertension.

PAH-targeted therapies are not generally used

Therapies specifically designed for pulmonary arterial hypertension (PAH), which is Group 1 pulmonary hypertension, have limited data supporting their use in Group 5 PH. They may be used on a case-by-case basis but have the potential to worsen oxygen levels, increase shortness of breath, and cause hypotension. A detailed discussion with your care team is required based on your circumstances.

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 health care team.
- 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 5 PH.

Pulmonary circulation image credit

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