Group 3 Pulmonary Hypertension

What is Group 3 pulmonary hypertension (PH due to lung disease and/or low oxygen)?

Group 3 pulmonary hypertension (Group 3 PH) is a type of pulmonary hypertension (high blood pressure in the lungs) that arises from **chronic lung diseases** or conditions that cause **low oxygen levels** (hypoxia).

What causes Group 3 pulmonary hypertension?

The most common causes include:

Chronic Obstructive Pulmonary Disease (COPD)

A group of lung diseases that block airflow.

Interstitial lung diseases (ILDs)

A group of lung diseases that cause scarring and thickening of the lung tissue. A common example is idiopathic pulmonary fibrosis (IPF).

Other lung diseases

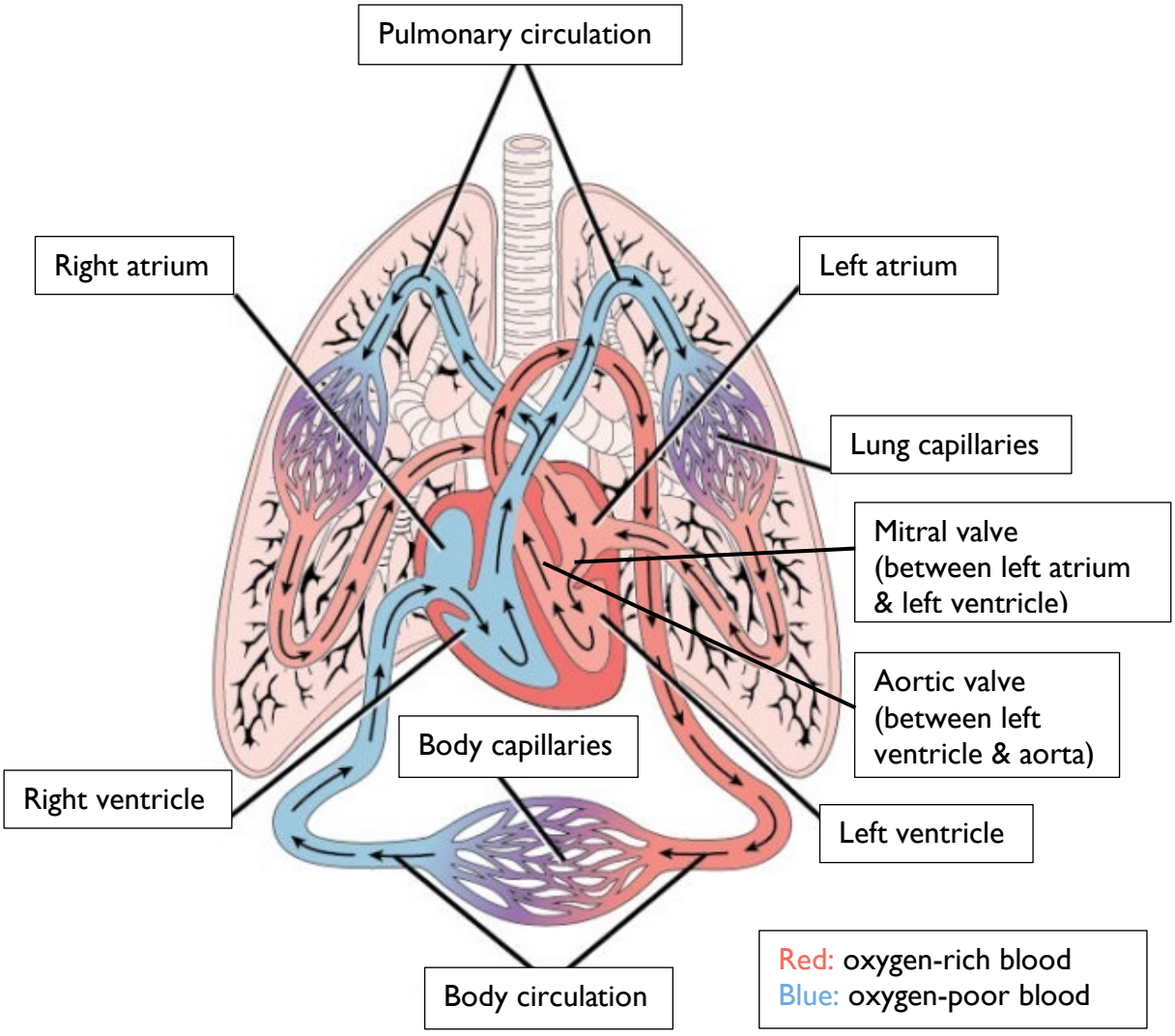
Conditions like combined pulmonary fibrosis and emphysema (CPFE), bronchiectasis, and cystic fibrosis.

Hypoxia (low oxygen levels)

Normal oxygen levels vary with age, but in healthy people, oxygen saturations are typically maintained above 94% at rest and above 90% on exertion. Values below this can cause or contribute to pulmonary hypertension. Examples include conditions such as obstructive sleep apnea, alveolar hypoventilation disorders, and chronic exposure to high altitudes.

What happens when you have Group 3 pulmonary hypertension?

In Group 3 PH, the number and/or width of small arteries in the lungs is reduced, leading to high blood pressure throughout the lungs, which then puts a progressive strain on the right side of the heart.



Group 3 PH is associated with increased shortness of breath and mortality (death) compared to people with lung disease who do not have 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

The primary goal of treatment is to manage the underlying lung disease and address any hypoxia, which can help improve the pulmonary hypertension.

Oxygen therapy

Patients with lung disease and pulmonary hypertension 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.

Other therapies

Other therapies depend on the specific condition.

- For COPD: Bronchodilators, inhaled corticosteroids, and other medications. Check with your team to ensure proper use of inhaled medications, as this is a common error.
- For interstitial lung diseases: Immunosuppression and antifibrotics. Inhaled treprostinil (Tyvaso) is not available in Canada, but is approved for the treatment of pulmonary hypertension related to ILD in the U.S.
- For sleep disordered breathing: Nocturnal Positive Pressure Therapy (e.g. CPAP or BPAP).
- For fluid retention: diuretics.

PH-targeted therapies are not generally used for this condition.

Therapies specifically designed for pulmonary arterial hypertension (PAH), which is Group 1 pulmonary hypertension, have limited data supporting their use in Group 3 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.

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 lung condition from your respirologist.
- 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 3 PH. Ongoing investigations are being conducted to improve outcomes in this condition.

Pulmonary circulation image credit

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