Group 2 Pulmonary Hypertension

What is Group 2 pulmonary hypertension (PH due to left heart disease)?

Group 2 pulmonary hypertension (Group 2 PH) is a type of pulmonary hypertension (high blood pressure in the lungs) that arises from **diseases of the left side of the heart**, including the left ventricle and the mitral and aortic valves.

What causes Group 2 pulmonary hypertension?

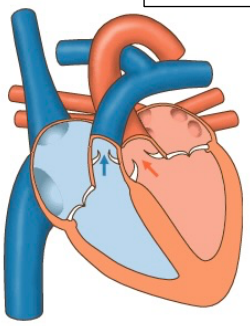
The most common causes include:

Heart failure

Both heart failure with reduced ejection fraction (**HFrEF**) and heart failure with non-reduced ejection fraction (**HFnrEF**) can contribute to Group 2 PH.

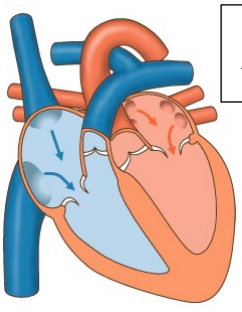
HFrEF occurs when the left side of the heart has a reduced ability to pump blood throughout the body. This can happen in coronary artery disease, among other conditions.

In **HFnrEF**, the left side of the heart pumps normally but it cannot fully relax between each heartbeat. Both lead to increased pressures in the pulmonary veins. Diabetes, hypertension and obesity increase the risk of HFnrEF.



Restrictive cardiomyopathy

When the heart muscle becomes stiff and unable to relax properly, it leads to impaired filling and increased pressure.

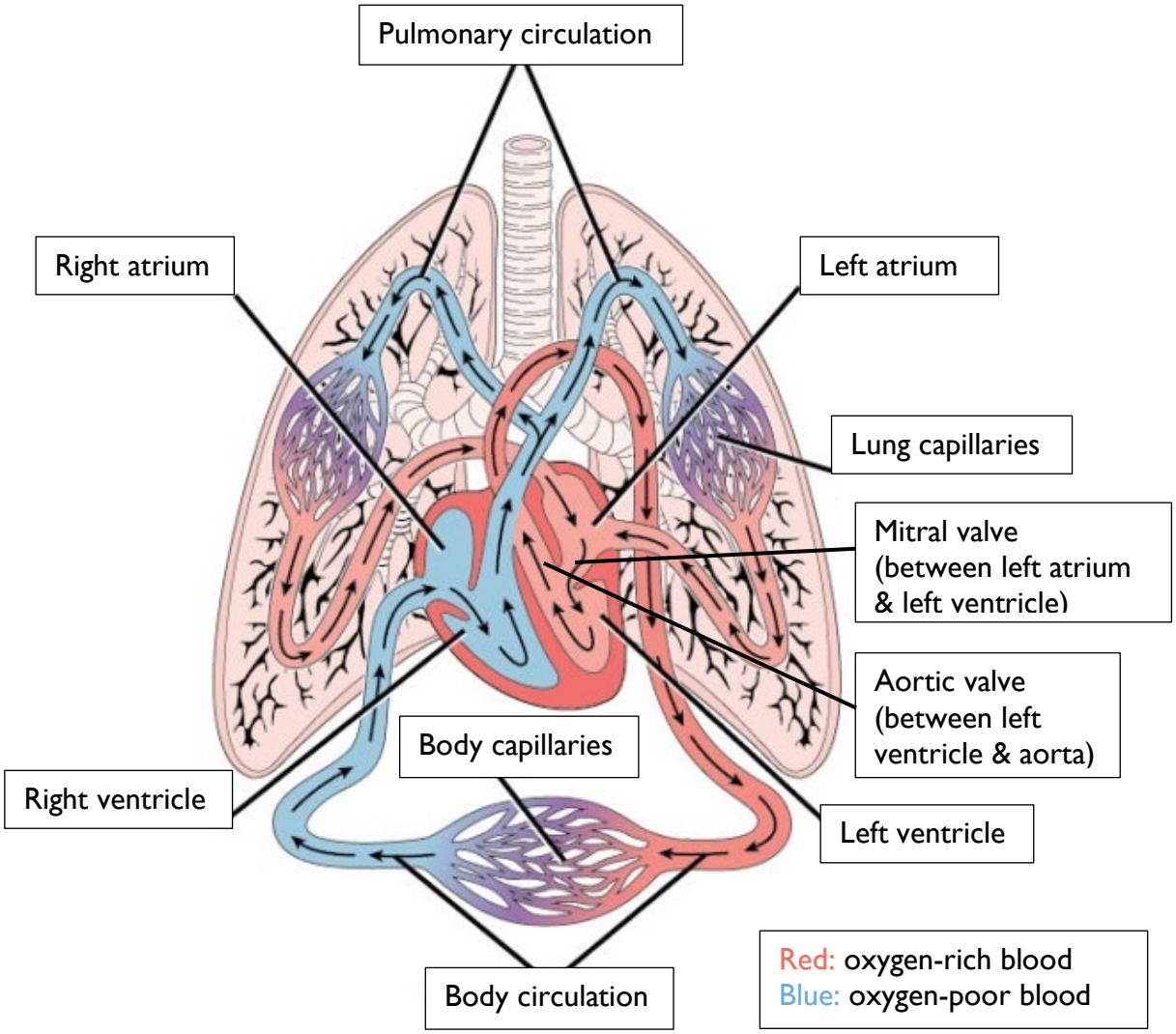


Valvular heart disease

Problems with the aortic and mitral valves can cause backflow of blood and pressure buildup. Valves may become leaky (regurgitation) or tight (stenosis).

What happens when you have Group 2 pulmonary hypertension?

When the left side of the heart isn't functioning properly, it can't effectively pump blood. This leads to a buildup of blood and pressure in the pulmonary veins, which are the veins that carry blood from the lungs back to the heart.



The increased pressure in the pulmonary veins is then transmitted further upstream in the pulmonary circulation to the pulmonary arteries, the blood vessels carrying the blood from the heart to the lungs. This causes the pressure to increase, resulting in pulmonary hypertension.

Group 2 PH can lead to impaired right ventricular function, worsening symptoms, and an increased risk of death.

Diagnosis

The diagnosis relies on a clinical probability assessment, with echocardiography playing a major role. Some patients require a procedure called a **right heart catheterization**, which will show elevation in the mean pulmonary artery pressure (mPAP), confirming pulmonary hypertension. Additionally, it measures the pulmonary artery wedge pressure (PAWP), which reflects elevated pressures in the left side of the heart.

Treatment

Treat the underlying heart disease

Your family physician and cardiologist will work together to manage and improve the underlying left heart disease. Common treatments include:

- Diuretics to relieve fluid retention
- Therapies directed at improving cardiac function (e.g. beta-blockers, ACE-inhibitors/angiotensin receptor blockers, angiotensin receptor/neprilysin inhibitors, SGLT-2 inhibitors, mineralocorticoid receptor antagonists, others)

No PH-targeted therapies are approved for this condition

Therapies specifically designed for pulmonary arterial hypertension (PAH), which is Group 1 PH, are generally not effective for Group 2 PH. Off-label use of medications like tadalafil, macitentan, and epoprostenol in Group 2 PH may lead to harm and is generally NOT recommended.

Other treatments

Some patients require other more advanced therapies, procedures, or surgery depending on the cause of their heart disease.

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 heart condition.
- Obtain a blood pressure cuff, heart rate monitor, scale and pulse oximeter. Learn how to monitor your vital signs and how to react to changes in weight.
- Watch for fluid retention and report it to your care team.
- 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.
- Ask your care team about any clinical trials in Group 2 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 used under a Creative Commons Attribution license

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