

PULMONARY ARTERIAL HYPERTENSION PATIENT INFORMATION LEAFLET

What is pulmonary arterial hypertension (PAH)?

Pulmonary arterial hypertension (PAH) is a progressive disease resulting from narrowing of lung arteries, which in turn limits blood flow to the lungs and results in high blood pressure in the lungs. Over time, this restriction in blood flow in the lung arteries increases the strain on the right heart to pump blood to the lungs. The right heart may become enlarged, resulting in right heart failure.

What are the symptoms?

The reduction in blood flow to the lungs, and in turn back to the left heart and main circulation, are the main reasons for symptoms. These symptoms include:

- Breathlessness
- Fatigue
- Dizziness especially during physical activity
- Chest discomfort or angina
- Temporary loss of consciousness or fainting
- Swollen ankles or legs due to fluid retention

Living with PAH

- 1. Reduce salt intake and take food labelled with “low-sodium”.**
- 2. Monitor fluid intake as recommended by your doctor.**
- 3. Stop smoking.**
- 4. Activity Guidelines**
 - Restrict lifting or pushing to less than 10kg, as these activities can increase pressure in your arteries and lungs.
 - Walking is important to keep your muscles strong and improve circulation. Strenuous activities which can lead to shortness of breath, giddiness or chest pain should be stopped immediately.
- 5. Avoid pregnancy, use 2 methods of birth control.**
 - Generally, recommend to use BOTH barrier type and oral hormone contraceptive pill (especially Progesterone-only pill)
- 6. Vaccinations**
 - Patients with PAH are prone to lung infections.

- Influenza and pneumococcal vaccinations are recommended.

7. Travelling



- May require oxygen at higher altitudes on the plane.

8. Over the counter medications

- Always ask your doctor or pharmacist before taking any over-the-counter medications.
- Some drugs such as **decongestant** (including Clarinase, Benadryl and other drugs containing ephedrine or pseudoephedrine) and **non-steroidal anti-inflammatory (NSAIDs)** agents such as Diclofenac (Voltaren) and Mefenamic Acid (Ponstan) can cause problems in people who have heart failure.
- Avoid any over-the-counter medications with labels which advise to use the medicine with caution if you have high blood pressure.

What are the treatment options?

i) Phosphodiesterase-5 inhibitor (PDE5i)

<p>Sildenafil</p> 		<p>Tadalafil</p> 			
How it works		Possible side effects		Note	
<p>Reduces breakdown of a protein (cGMP) which relaxes blood vessels in lung, thus reduces lung blood pressure.</p>		<p>Headache, flushing and skin redness, dizziness, upset stomach, stuffy nose, insomnia, low blood pressure, ringing of the ears.</p>		<p>DO NOT take with Riociguat (Adempas).</p> <p>DO NOT take with Nitrates eg: isosorbide mononitrate (ISMN), isosorbide dinitrate (ISDN), glyceryl trinitrate (GTN), as the combination of these medicines may lead to a dangerous fall in your blood pressure. Always tell your doctor or pharmacist if you are taking any of these medicines that are often used for the relief of angina pectoris (or “chest pain”).</p>	




ii) Soluble Guanylate Cyclase (sGC) Stimulator

Riociguat



How it works	Possible side effects	Note
Increases production of a protein (cGMP) which relaxes blood vessels in lung, thus reduces lung blood pressure.	Low blood pressure (dizziness), headache, nausea, diarrhoea, upset stomach.	<ul style="list-style-type: none"> - Smoking may reduce effectiveness of treatment - Avoid taking with antacids (e.g. Gaviscon) - Do not take this medicine if you are pregnant or plan to become pregnant. - DO NOT take with Nitrates eg: isosorbide mononitrate (ISMN), isosorbide dinitrate (ISDN), glyceryl trinitrate (GTN), or PDE5I eg: Sildenafil (Viagra), Tadalafil (Cialis) as this can result in a dangerous fall in your blood pressure.

iii) Endothelin receptor antagonists (ERA)

<u>Bosentan</u>	<u>Macitentan</u>	<u>Ambrisentan</u>
		
How it works	Possible side effects	Note
Blocks the action of a substance (Endothelin) which cause narrowing of lung blood vessels, thus reduces lung blood pressure	<p>Bosentan: Headache, dizziness, flushing, liver abnormalities, water retention and leg swelling, irregular heartbeats.</p> <p>Macitentan: Cold-like symptoms, Low red blood count (anaemia), headache.</p> <p>Ambrisentan: Leg swelling, headache, nasal congestion, low red blood count</p>	<ul style="list-style-type: none"> - Do not take this medicine if you are pregnant or plan to become pregnant. - Reduces effectiveness of oral contraceptive pill - Monitor for yellowing of eye white, skin, or dark tea-coloured urine. - Monitor red blood count after first month of therapy then periodically as advised (for macitentan and ambrisentan). <p>For Bosentan:</p> <ul style="list-style-type: none"> - Requires liver test once monthly. - May significantly reduce the effectiveness of Warfarin. INR should be monitored more

		frequently when bosentan is initiated, adjusted or discontinued in patients taking warfarin.
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iv) Prostacyclin

(a) Inhaled

Iloprost inhalation solution



How it works	Possible side effects	Note
Directly helps to activate relaxation of lung blood vessels, thus reduces lung blood pressure	Cough, flushing, headache, nausea/vomiting, jaw and muscle pain, low blood pressure (dizziness), flu-like symptoms, risk of infection and plug site blood clotting.	Need nebulizer. A specialized nurse will train caregiver on usage at home

(b) Intravenous:

Examples: Iloprost (ilomedin), Epoprostenol (Flolan)
For more advanced disease and only in the hospital setting

Supportive treatments

The need to start these medicines depends on physician assessment

Therapy	Medicine	When it is needed	Note
Oxygen	Inhaled oxygen	When blood oxygen is too low due to shortness of breath	Requires a machine and inhaled through a nose canal or face mask

Diuretics ("Water pill")	Frusemide Bumetanide Metolazone	This medicine helps to get rid of excess fluid in the body e.g. leg swelling or when water in lungs cause shortness of breath.	Will have increased urination
Blood thinner	Warfarin	When the risk of developing blood clots in the body is high	Nil