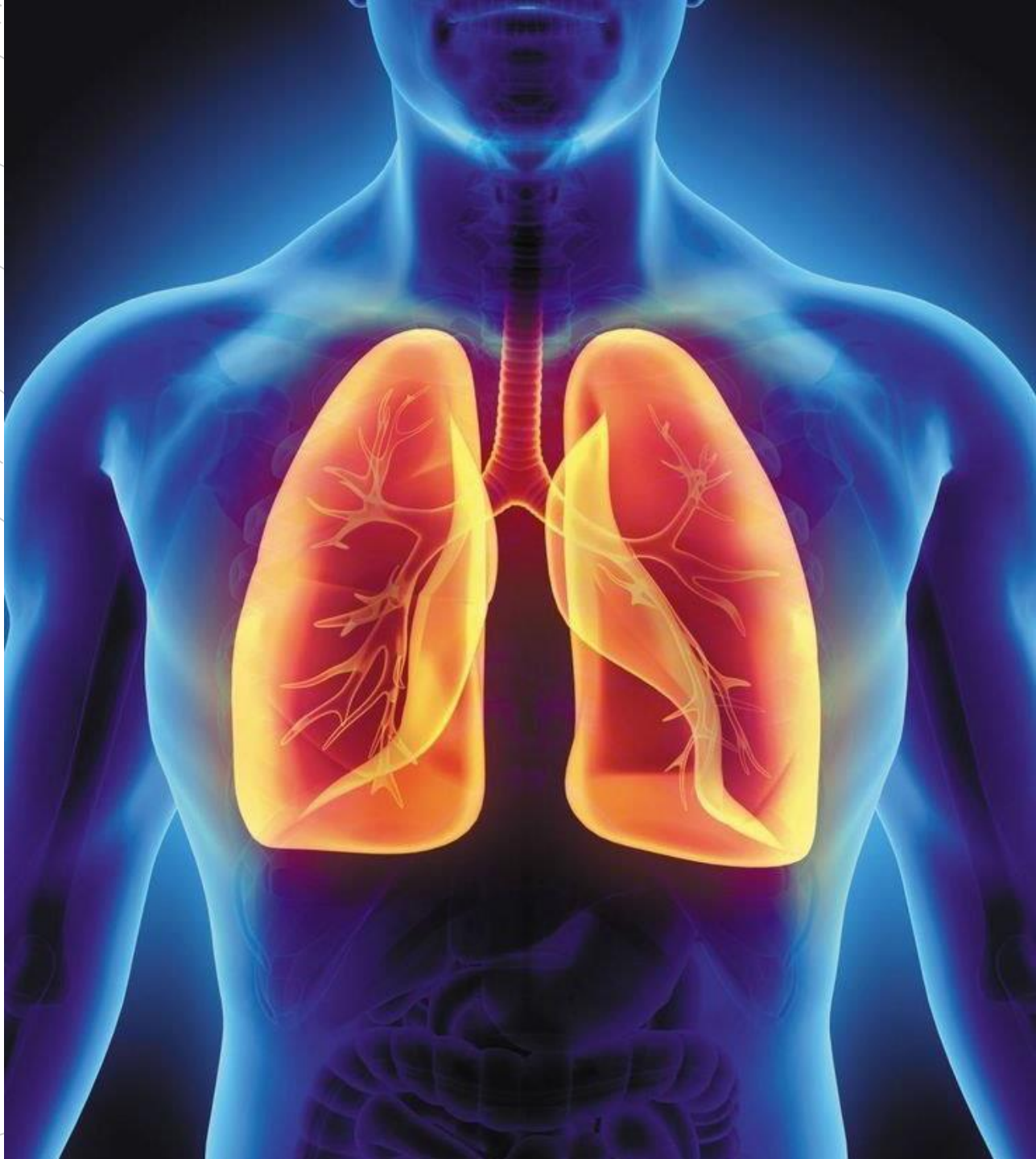
Pulmonary hypertension

Rama Salim

Yaqeen Farajat



Definition

- pulmonary hypertension is a condition that affects the blood vessels in the lungs. It develops when the blood pressure in your lungs is higher than normal.
- It happens when the mean pulmonary artery pressure (PAP) is at least 25 mmHg at rest, as measured by right heart catheterisation .
- mPAP = 2/3 dPAP + 1/3 sPAP , where dPAP is diastolic pulmonary artery pressure, and sPAP is systolic pulmonary artery pressure

Pathological features

Pathological features include:
1- hypertrophy of both the media
and the intima of the vessel wall and a clonal expansion of endothelial cells, which take on the appearance of plexiform lesions.
2-There is marked narrowing of the vessel lumen and this, together with the frequently observed in situ thrombosis, leads to an increase in pulmonary vascular resistance and PH

Classification of pulmonary hypertension

- Pulmonary hypertension can develop on its own or be caused by another disease or condition.
- The clinical classification is based on the degree of functional disturbance .
- Although respiratory failure due to intrinsic pulmonary disease is the most common cause of PH.

There are five different groups of pulmonary hypertension:

1-Pulmonary arterial hypertension

2- Pulmonary venous hypertension

3- Pulmonary hypertension associated with disorders of the respiratory system and/or hypoxaemia

4-Pulmonary hypertension caused by chronic thromboembolic disease

5-Miscellaneous

Pulmonary arterial hypertension

1) Primary pulmonary hypertension: sporadic and familial

- ✓ Primary pulmonary hypertension (PPH) is a rare but important disease that predominantly affects women aged between 20 and 30 years.
- ✓ Familial disease is rarer still but is known to be associated with mutations in the gene encoding type II bone morphogenetic protein receptor (BMPR2), a member of the transforming growth factor beta (TGF-β) superfamily.
- ✓ Mutations in this gene have been identified in some patients with sporadic PH
- ✓ symptoms:

1. Asymptomatic in mild hypertension
2. Exertional dyspnea is the most common presentation
3. Dizziness, syncope and chest pain in severe cases (poor prognosis).

Pulmonary arterial hypertension

✓ Signs:

1) Prominent pulmonic component (P2) of the S2 is a reliable indicator of elevated PA pressure

2) Added sound: 3, holosystolic murmur (tricuspid regurgitation)

3) Parasternal heave (right ventricular hypertrophy) & High JVP

✓

✓ The prognosis is poor . Mean survival is 2-3 years from the time of diagnosis .

2) Secondary to: connective tissue disease (limited cutaneous systemic sclerosis), congenital systemic to pulmonary shunts, portal hypertension, HIV infection, exposure to various drugs or toxins, and persistent pulmonary hypertension of the newborn

Pulmonary venous hypertension

1) • Left-sided atrial or ventricular heart disease

2) • Left-sided valvular heart disease

3) • Pulmonary veno-occlusive disease

4) • Pulmonary capillary haemangiomatosis

Pulmonary hypertension associated with disorders of the respiratory system and/or hypoxaemia

1)• Chronic obstructive pulmonary disease
2)• Diffuse parenchymal lung disease
3)• Sleep-disordered breathing
4)• Alveolar hypoventilation disorders
5)• Chronic exposure to high altitude
6)• Neonatal lung disease
7)• Alveolar capillary dysplasia
8)• Severe kyphoscoliosis

pulmonary hypertension caused by chronic thromboembolic disease

1) • Thromboembolic obstruction of the proximal pulmonary arteries
2) • In situ thrombosis
3) • Sickle-cell disease

Miscellaneous

1)• Inflammatory conditions

2)• Extrinsic compression of central pulmonary veins

Clinical features

PH presents insidiously and is often diagnosed late. Typical **symptoms** include:

1- breathlessness

2- chest pain

3-fatigue

4- palpitation

5- syncope (with sever disease)

- Important **signs** include elevation of the JVP (with a prominent 'a' wave if in sinus rhythm),
- a parasternal heave (right ventricular hypertrophy),
- accentuation of the pulmonary component of the second heart sound and a right ventricular third heart sound.
- Signs of interstitial lung disease or cardiac, liver or connective tissue disease may suggest the underlying cause.

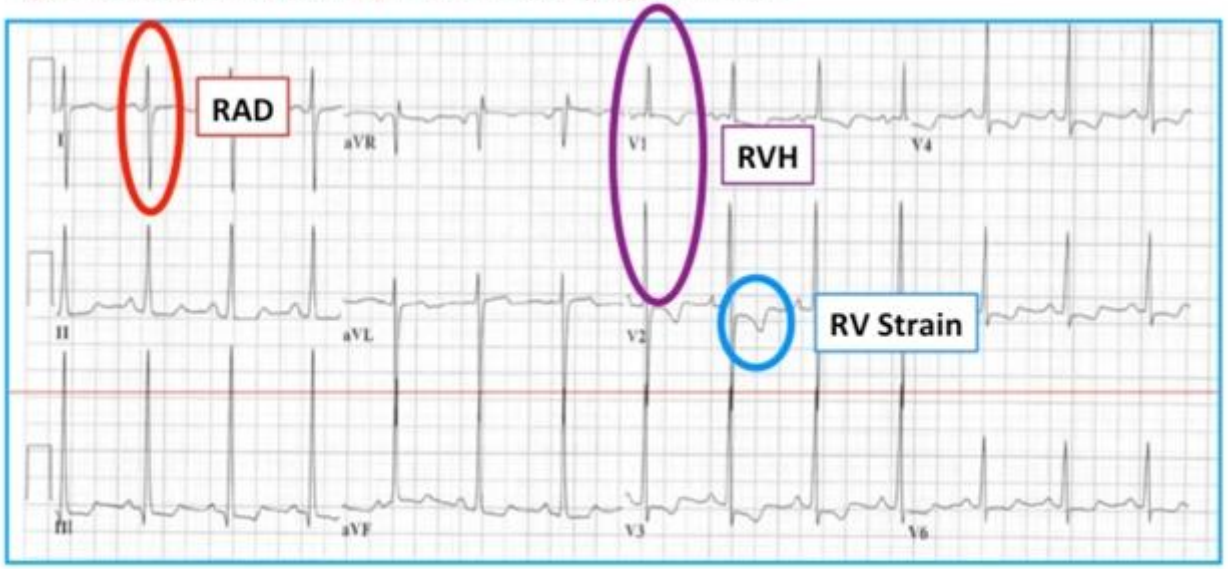
-The height of the jugular venous pulse is determined by right atrial pressure and is therefore elevated in right heart failure and reduced in hypovolaemia.
- The a wave represent the atrial contraction

Investigations

ECG

- ECG often shows right ventricular strain(ST depression and T wave inversion in leads V1 to V3).
- Other changes found include right axis deviation and right atrial abnormality

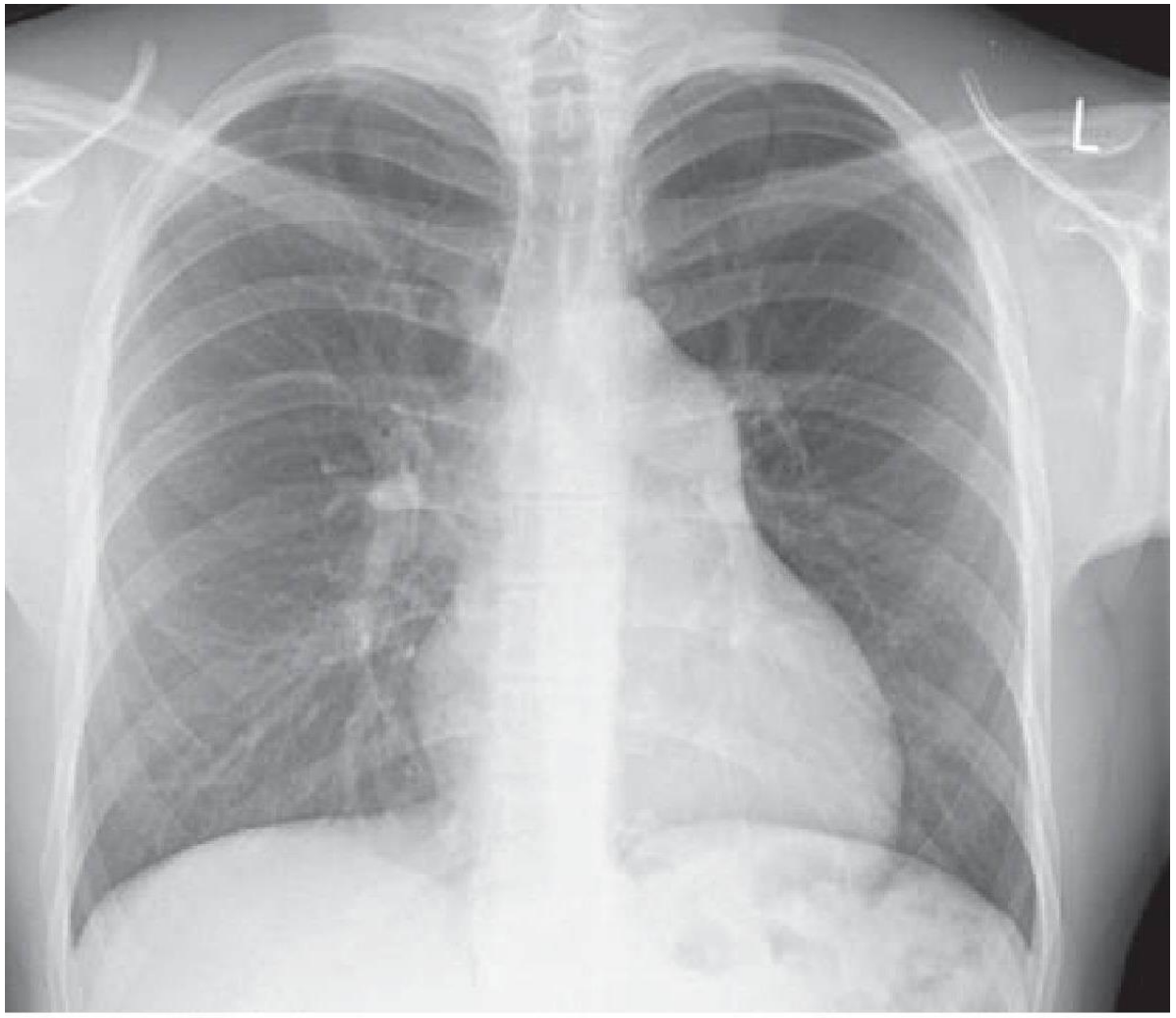
Figure 1. Sample ECG with Signs of Pulmonary Hypertension



PAH, pulmonary arterial hypertension; RAD, right axis deviation; RVH, right ventricular hypertrophy; RV, right ventricle.

Chest x-ray

- Enlarged pulmonary arteries with rapid tapering of vessels toward the periphery of the lungs (a "pruned tree" appearance)
- Right-heart enlargement

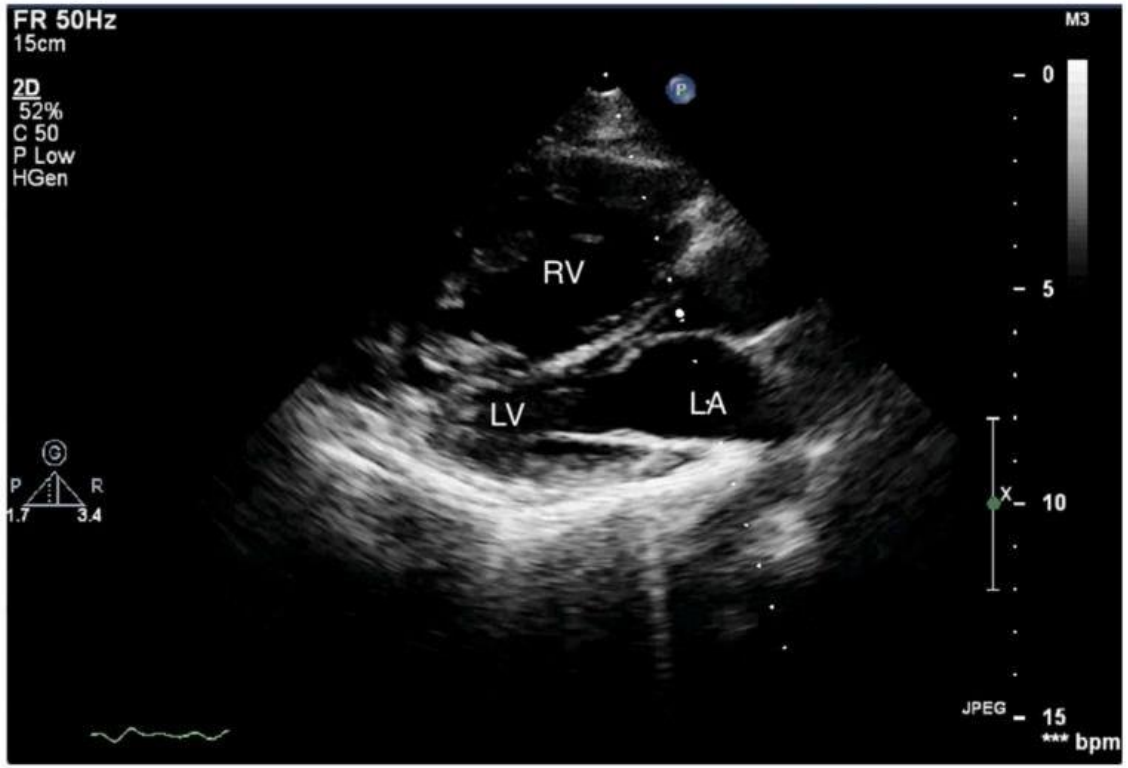


Chest X-ray showing the typical appearance in Pulmonary hypertension.

Echocardiography

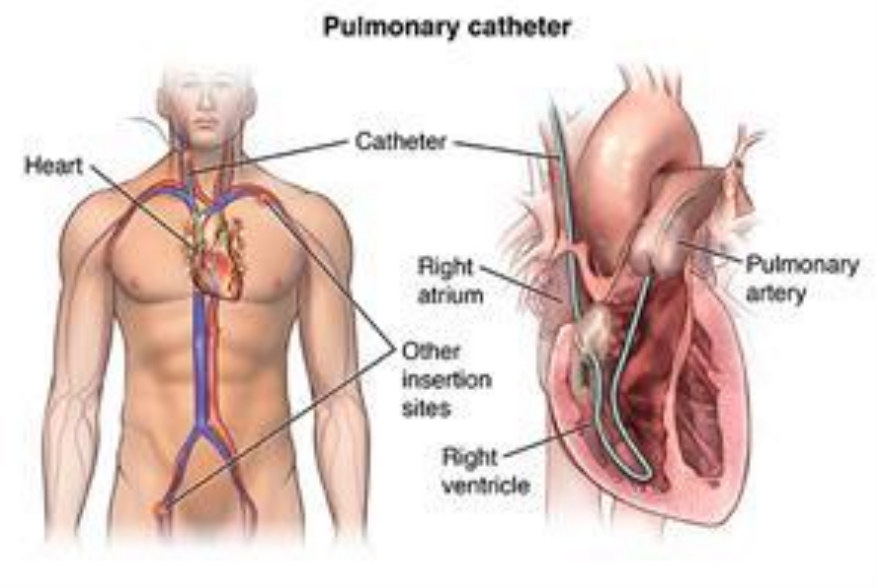
- Signs of right ventricular pressure overload:

1. Paradoxical bulging of the septum into the left ventricle during systole.
2. Dilation /hypertrophy of the right ventricular free wall
3. Dilation /hypertrophy of RA



Right-heart catheterization

Permits direct measurement of Pulmonary artery pressure, and with angiography, a definitive diagnosis of chronic thromboembolic disease.



Management

Treatments for pulmonary hypertension will depend on the cause of the condition. Many times, there is no cure for pulmonary hypertension, but your provider can work with you to manage the symptoms. This may include medicine or healthy lifestyle

- Diuretic therapy should be prescribed for patients with right heart failure.
- Supplemental oxygen should be given to maintain resting PaO2 above 8 kPa (60 mmHg).
- Anticoagulation should be considered unless there is an increased risk of bleeding.
- Digoxin may be useful in patients who develop atrial tachyarrhythmias.
- Pregnancy carries a very high risk of death and women of child-bearing age should be counselled appropriately.
- Excessive physical activity that leads to distressing symptoms should be avoided but otherwise patients should be encouraged to remain active
- Drugs should be avoided :

1-Nitrates ; owning the risk of hypotension

2- β-blockers ;are poorly tolerated

3-Cyclizine can aggravate PH and should also be avoided.

- Disease-targeted strategies :

1. they have focused on replacing endogenous prostacyclins with infusions of epoprostenol or treprostinil or nebulised iloprost
2. blocking endothelin-mediated vasoconstriction with agents such as bosentan, ambrisentan or macitentan
3. enhancing endogenous nitric oxide-mediated vasodilatation with phosphodiesterase V inhibitors, such as sildenafil or tadalafil, or the guanylate cyclase stimulator riociguat.

- **High-dose calcium channel blockers may be appropriate in those with an acute vasodilator response**
- Selected patients are referred for double-lung transplantation, and pulmonary thrombo-endarterectomy may be contemplated in those with chronic proximal pulmonary thromboembolic disease

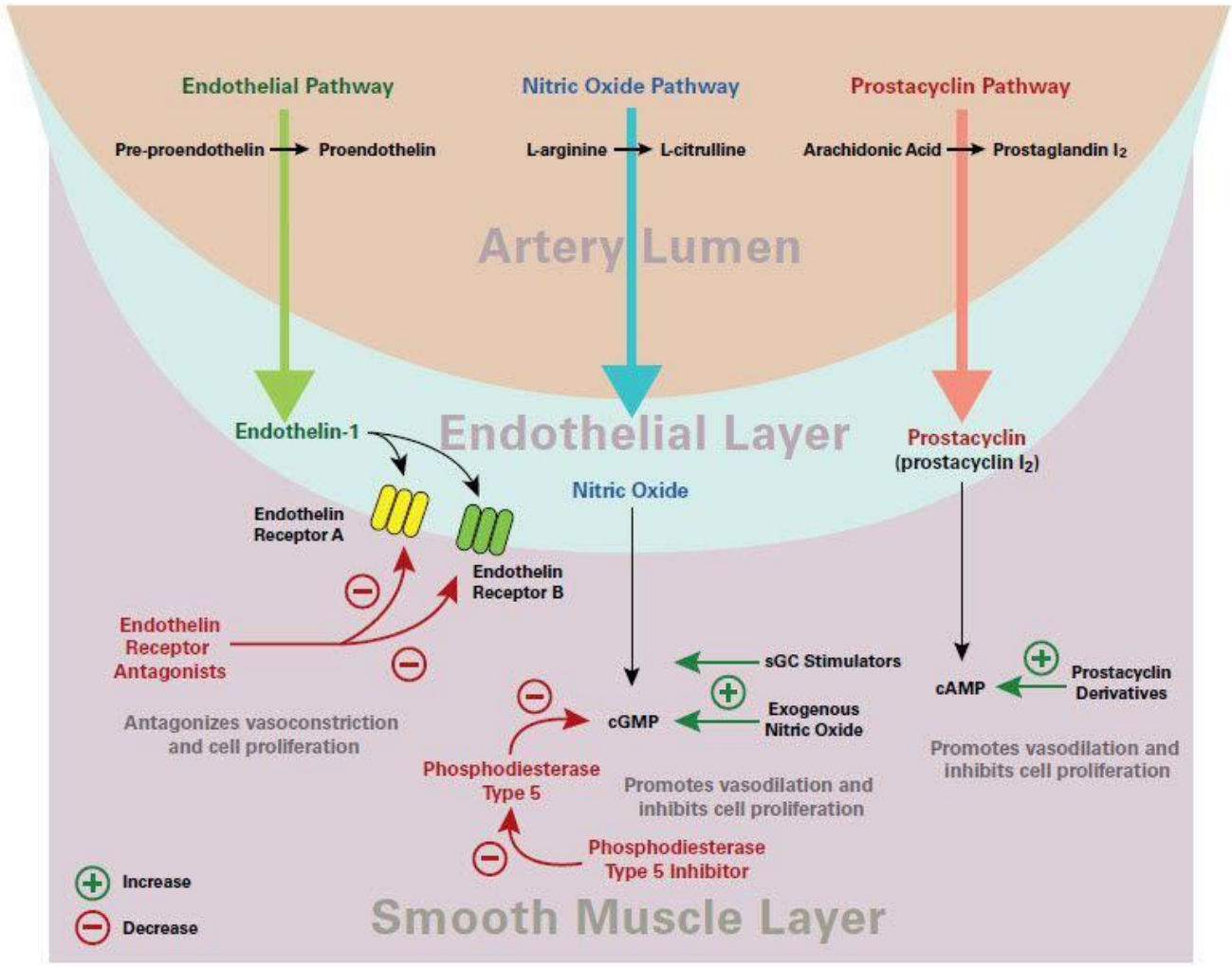


Figure 1. Target Pathways and Current Therapies in Pulmonary Arterial Hypertension[17,18]

cAMP indicates cyclic adenosine monophosphate; cGMP, cyclic guanosine monophosphate; sGC, soluble guanine cyclase.
Schematic diagram of 3 biologic pathways involved in the pathogenesis of pulmonary arterial hypertension.
Adapted from Humbert et al. *N Engl J Med.* 2004;351(14):1425-1436.

Thank you

source : Davidson's Principles and Practice of Medicine