Pulmonary Arterial Hypertension

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INTRODUCTION

Pulmonary Arterial Hypertension (PAH), also referred to as WHO Group 1 pulmonary hypertension, is a chronic, rare, and progressive disease characterized by high blood pressure in the arteries of the lungs, specifically with a mean pulmonary artery pressure of >20 mmHg.

The incidence of pulmonary arterial hypertension (PAH) has increased significantly over the past three decades, with global incident cases rising by 85.6% from 1990 to 2021.
> Only about 1000 new cases of PAH are diagnosed every year in the United States.
> PAH is more common in women than in men.
> PAH is chronic, and it gets worse over time.
> Treatment options and approaches to care, such as improving the risk status, continue to advance.

METHODS

Although PAH is rare, it is often fatal in the long term; hence, we decided to evaluate its pathophysiology through a literature search.

In PAH, blood vessels in the lungs are narrowed, blocked, or destroyed (Figures 1, 2). The damage makes it hard for blood to move through the lungs. Blood pressure in the pulmonary arteries rises. The heart must work harder to pump blood through the lungs. The extra effort eventually causes the heart muscle to become weak and fail.

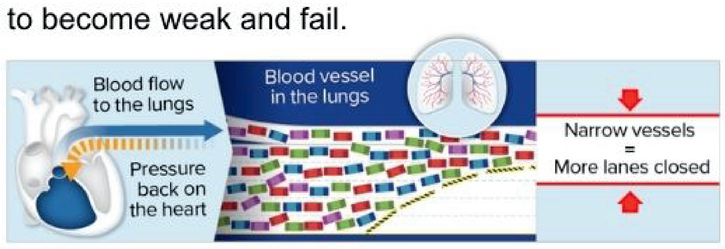

Figure 1: Pathophysiology of PAH showing narrowed pulmonary vessels and increased pressure on the heart

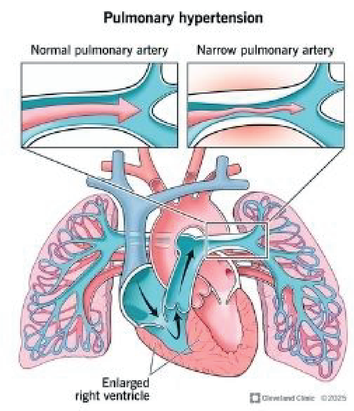

Figure 2: Comparison of normal and narrowed pulmonary arteries in PAH

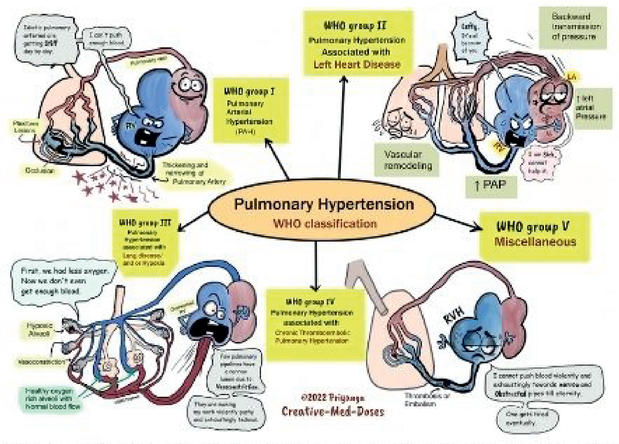

Figure 3: WHO Functional classification groups of PAH

METHODS, CONTD.

Stages of pulmonary hypertension
There are four main stages or "functional classes" (Figure 3). As PAH gets worse, the symptoms become more obvious and disruptive (Figure 4).

- Class 1: No symptoms.
- Class 2: Mild symptoms. The individual feels fine while resting.
- Class 3: Moderate symptoms. It's much harder to do normal tasks, feeling very tired or short of breath, but still feel fine when resting.
- Class 4: Severe symptoms, even at rest. The symptoms worsen with mobility.

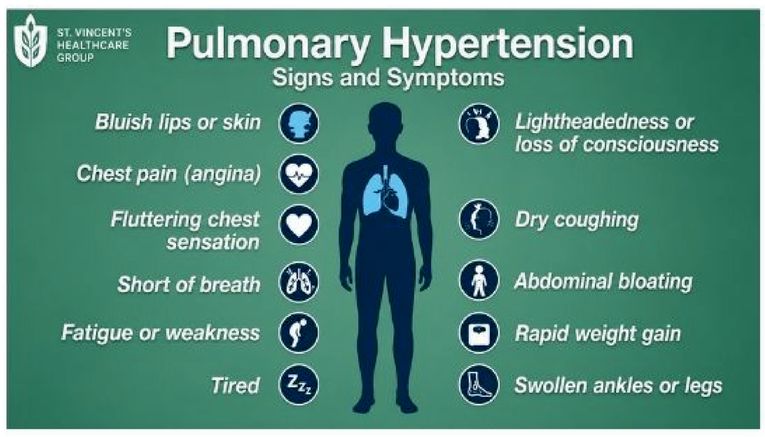

Figure 4: Clinical presentation of PAH

RESULTS

Diagnosis

- **Physical Exam,** including vital signs.
- **Blood tests** to check for issues related to organ function, hormone levels, and infections.
- **Six-minute walk test.**
- **Chest X-ray.**
- **Doppler echocardiogram**.
- **Chest CT scan**.
- **Right heart catheterization**.
- **V/Q (Ventilation-Perfusion) scan.**

Management

Most cases of pulmonary hypertension cannot be cured. The various treatment modalities include:

- **Pulmonary vasodilators** (Figure 5).
- **Oxygen therapy.**
- Groups 2 and 3: Medications (Adempas), dietary changes, oxygen therapy, or surgery.
- Group 4: **Balloon pulmonary angioplasty.**
- **Pulmonary endarterectomy**.
- Group 5: **Lung transplant.**

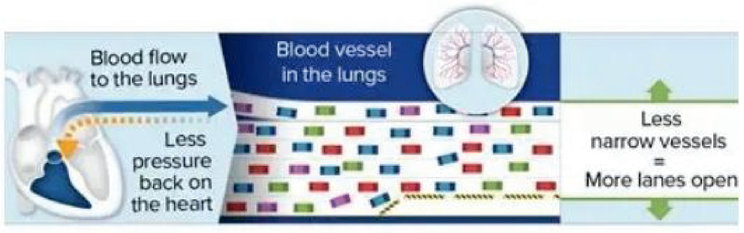

Figure 5: Blood flow after treatment of PAH using pulmonary vasodilator

RESULTS, CONTD.

- Anemia.
- Abnormal heart rhythms.
- Blood clots in your pulmonary arteries
- Pericardial effusion.
- Pregnancy complications (PAH is dangerous for both the mother and the fetus).
- Right-sided heart failure (Figure 6).

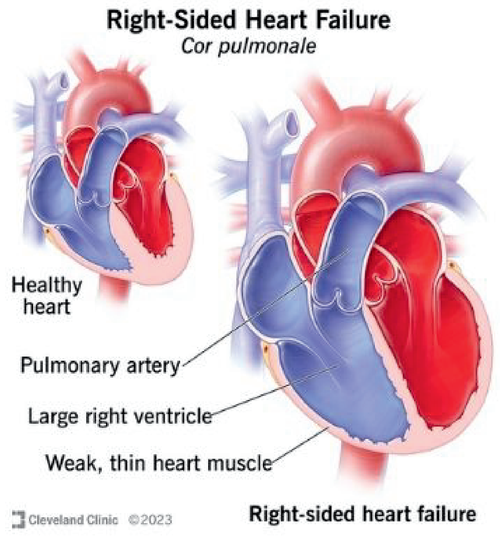

Figure 6: Comparison of healthy heart and enlarged heart in right-sided heart failure.

Life expectancy of a patient with PAH

- The life expectancy of someone with PAH. depends on how early the disease is diagnosed and how well the patient responds to treatment.
- With modern treatments, life expectancy has increased by 10 years or more for some patients.

CONCLUSIONS

- PAH is a rare but serious and progressive condition that leads to right heart failure and ultimately death.
- PAH can be classified into several groups and stages.
- Diagnosis involves a comprehensive patient evaluation, including invasive right heart catheterization.
- The life expectancy in PAH depends on how early the condition is diagnosed and how well the patient responds to treatment. With modern treatments being introduced, patients are able to live much longer.
- Although there is no cure, treatments such as oxygen therapy, medications like Adempas, and surgical procedures can help manage the symptoms of PAH, slow PAH progression, and improve overall quality of life.
- In summary, understanding the pathophysiology, diagnosis, prognosis, and management of PAH is vital, as early recognition and targeted interventions can prevent irreversible right heart failure and significantly improve survival and quality of life.