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# Pulmonary Arterial Hypertension

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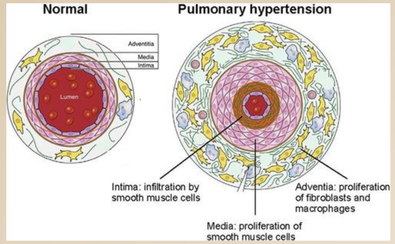
## Introduction

- Pulmonary arterial hypertension (PAH) is characterized by persistently elevated pressures in the pulmonary arteries (Poch, 2016).
- PAH is a rare disease with about 500 - 1000 new cases a year in the U.S. The prevalence of PAH is equal among all ethnicities and races (Raredisease.org, 2020).
- PAH can be life-threatening with a general life expectancy of 5 years if left untreated or treated inadequately, making early diagnosis and treatment crucial for patients and healthcare providers (Poch, 2016).
- As a former cardiac nurse who has taken care of PAH patients, and witnessed how devastating the disease is even with treatment, I feel the need to bring awareness to early diagnosis and treatment.

## Diagnosis

- PAH can be challenging to diagnose due to non-specific presenting symptoms (Ricketts & Church, 2017).
- The average time between the onset of symptoms and diagnosis of PAH ranges anywhere from 18-32 months (Prins & Thenappan, 2016).
- Right heart catheterization is regarded as the gold standard for PAH diagnosis as it provides the hemodynamic data that defines the disease (Pristera, Musarra, Schilz, & Hoit, 2015).
- Echocardiography, although not considered the gold standard for the diagnosis of PAH, can be used to diagnose PAH. Echocardiography is also used to assess the existence of right ventricular (RV) or right atrial enlargement or dilation and cardiac causes of PAH, such as valvular heart disease, left ventricular dysfunction, or pericardial effusion (Leeper & Powell, 2019).
- PAH is defined hemodynamically as a mean pulmonary arterial pressure (mPAP) greater than or equal to 25 mm Hg at rest (Pristera, Musarra, Schilz, & Hoit, 2015).
- There are 5 types of pulmonary hypertension (PH), according to the World Health Organization. They are (1) PAH, (2) PH due to left heart disease, (3) PH due to lung disease, or hypoxia, (4) chronic thromboembolic PH, and (5) PH with unclear multifactorial means (Pristera, Musarra, Schilz, & Hoit, 2015).
- The WHO recognizes 4 functional classes in PH. Functional class I is considered the mildest form of the disease, and class IV is the most severe ( Ricketts & Church, 2017).




(Figure1 Sysol & Machado, 2018)

## Signs and Symptoms

- PAH can be challenging to diagnose due to non-specific signs and symptoms initially, but the following are the symptoms patients usually present with (Ricketts & Church, 2017).
- Lethargy
- Malaise
- Exercise intolerance
- Dyspnea
- Exertional syncope and angina is seen as the disease progresses
- Development of Tricuspid regurgitation
- Increased intensity of the second heart sound
- Peripheral edema, ascites, and elevated jugular venous pressure can develop when the right heart chambers hypertrophy, and the left ventricle fails.
- Pulsatile hepatomegaly

| Functional class | Symptomatic profile |
|---|---|
| I | Patients with pulmonary hypertension but without resulting limitation of physical activity. Ordinary physical activity does not cause dyspnea or fatigue, chest pain, or near syncope |
| II | Patients with pulmonary hypertension resulting in slight limitation of physical activity. They are comfortable at rest. Ordinary physical activity causes undue dyspnea or fatigue, chest pain, or near syncope |
| III | Patients with pulmonary hypertension resulting in marked limitation of physical activity. They are comfortable at rest. Less than ordinary activity causes undue dyspnea or fatigue, chest pain, or near syncope |
| IV | Patients with pulmonary hypertension with inability to carry out any physical activity without symptoms. These patients manifest signs of right heart failure. Dyspnea and/or fatigue may even be present at rest. Discomfort is increased by any physical activity |

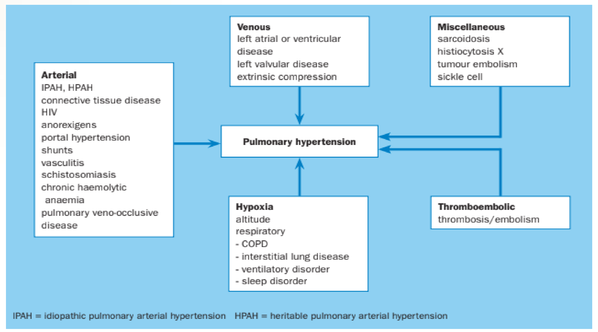
(Table 1. WHO functional classification of pulmonary arterial hypertension. (Ricketts & Church, 2017)

## Underlying Pathophysiology

- PAH is a disease that mostly affects the small resistance pulmonary arteries shown by intimal hyperplasia, medial hypertrophy, adventitial proliferation, localized thrombosis, and inflammation (Prins & Thenappan, 2016).
- PAH can be idiopathic, hereditable, drug and toxin-induced, or associated with connective tissue diseases, HIV infections, portal hypertension, congenital heart disease, and schistosomiasis (Prins & Thenappan, 2016).
- The gene involved in the hereditary PAH is the BMPR2 gene. The BMPR2 gene controls the number of cells in certain tissues. Mutation in the gene increases cell division or cell death leading to excessive growth of cells in the small arteries in the lungs (NIH, 2017).
- The excessive growth from the BMPR2 gene mutations narrows the small pulmonary arteries' diameter, which leads to resistance. To compensate for the resistance, BP in the pulmonary artery and the heart's right ventricle go up to overcome the increased resistance (NIH, 2020).
- There is persistent elevation in the right ventricle (RV) afterload as a result of pulmonary vascular resistance (PVR), which causes right ventricular hypertrophy (Prins & Thenappan, 2016).
- The disease process of PAH includes pulmonary vascular dysfunction associated with an imbalance of vasoconstricting and vasodilating substances in the small pulmonary arteries. The imbalance has been shown to favor the vasoconstricting activity, which causes the small pulmonary arteries to constrict ( Prins & Thenappan, 2016).

## Significance of Pathophysiology

- PAH is a complex disease that requires expert care (Ricketts & Church, 2017).
- The condition ends up affecting all the major organs of the body. The main issue with PAH is right-sided heart failure which then causes a more complicated clinical syndrome that affects multi-organ systems such as the left heart, brain, kidney, liver, gastrointestinal tract, skeletal muscle, endocrine, immune, and autonomic systems (Rosankranz, Howard, Gomberg-Maitland, & Hoeper, 2020).




(Figure 2. Classification of pulmonary hypertension, WHO 2008 (modified and not exhaustive)1 (Ricketts & Church, 2017).

## Implications for Nursing Care

- Nursing play a huge role in educating patients and family about the condition. Newly diagnosed PAH patients need information and education regarding the disease tailored to them and given at the right time (Graarup, Ferrari, & Howard, 2016).
- Nurses who take care of PAH patients need to be educated on the disease and current therapies. The focus should be on proper monitoring of patients receiving treatment with continuous IV therapies such as Epoprostenol due to the short half-life of the drug (Smith, York, Kane, & Weitendorf, 2015).
- Always have a backup IV pump, and an extra drug on the unit for pump malfunction or drug disconnections as an interruption in the drug's flow can cause life-threatening rebound elevation in pulmonary vascular resistance that can lead to acute heart failure and death (Ricketts & Church, 2017).
- Nurses caring for PAH patients need to monitor for complications such as line infections, thrombosis, and embolism (Ricketts &Church, 2017).
- Make sure that patients are eating their meals as nutritional deficiencies have been seen with PAH (Vinke, Jansen, Witkamp, & Norren, 2018).
- Nurses caring for PAH patients need to monitor them for signs and symptoms of RV failure (Leeper & Powell, 2019).
- Monitor oxygenation levels and start oxygen therapy for a Pao2 of less than 60 mmHg (Leeper & Powell, 2019).
- The nurse must report adverse medication reactions to the provider and give as-needed medications to reduce adverse reactions like nausea and diarrhea (Leeper & Powell, 2019).
- Nurses caring for PAH patients need to do a full head-to-toe assessment and report any changes to providers (Leeper & Powell, 2019).

## Conclusions

- PAH is a life-threatening disease with a general life expectancy of 5 years if left untreated or treated inadequately, making early diagnosis and treatment crucial for patients and healthcare providers (Poch, 2016).
- PAH disease-targeted therapy can only be started by doctors in specialist centers who specialize in treating PAH (Ricketts & Church, 2017).
- The initial signs and symptoms can be vague, which delays prompt diagnosis (Ricketts & Church, 2017).

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