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Pathophysiology of Pulmonary Hypertension

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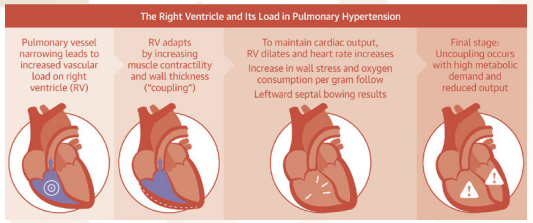
Introduction

- Pulmonary hypertension (PH) is a collection of related processes leading to vasoconstriction, vascular remodeling, endothelial dysfunction, and thrombosis (Sarkar & Desai, 2018).
- Those with PH have a mean pulmonary arterial pressure (mPAP) of >25 mmHg at rest and > 30mmHg during exercise (Sarkar & Desai, 2018).
- There are five types of PH (Sarkar & Desai, 2018).
- PH affects up to fifty million people and can lead to death, especially considering delayed diagnosis (Wood et al., 2021).
- Misdiagnosis is an ongoing problem for those with PH, leading to severe consequences (Kim & George, 2019).
- Life span of those without treatment is 2.8 years (Kim & George, 2019).
- Prognosis for those with PH is poor, with 1-year survival rates at 90.4% and 5-year survival rates at 65.4% (Bandyopadhyay et al., 2021).
- Topic selection stemmed from the importance of understanding PH pathophysiology for healthcare professionals, specifically nurses and nurse anesthetists, for safe delivery of care and anesthesia (Wood et al., 2021).
- As treatment improves for PH and patients live longer, an increase in surgical and medical emergencies resulting from PH co-morbidities emphasizes the importance of understanding the disease (Price et al., 2021).
- Those with PH undergoing surgery are at increased risk of complications resulting in death, further supporting the topic's importance (Price et al., 2021).



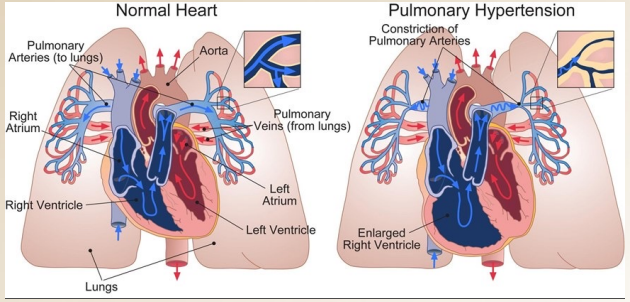
(Noordegraaf et al., 2017)

Signs & Symptoms

- Signs and symptoms of PH are often experienced for an extended period before diagnosis, with 20% of patients reporting symptom onset more than two years prior to diagnosis (Poch & Mandel, 2021).
- Progressive dyspnea is present in 85% of patients with PH, and over half claim it as their initial symptom (Poch & Mandel, 2021).
- Additional non-specific symptoms include dyspnea, lethargy, and palpitations resulting from hypoxemia (Wood et al., 2021).
- Over time patients may present with fluid overload leading to peripheral edema, ascites, and exertional chest pain (Sarkar & Desai, 2018; Wood et al., 2021).
- Exertional syncope resulting from decreased cardiac output and additional oxygen demand is a final symptom of PH (Wood et al., 2021).
- A common and extreme sign of PH is right ventricular (RV) uncoupling with the pulmonary circulation over time (Price et al., 2021).
- The RV initially attempts to undergo hypertrophy to augment stroke volume, but due to disease advancement eventually dilates with an increase in heart rate resulting in the uncoupling (Noordegraaf et al., 2017).
- Common physical findings include elevated jugular venous pressure, right-sided additional heart sounds, tricuspid regurgitation, and hepatomegaly (Kondo et al., 2019; Poch & Mandel, 2021).
- Syncope is an important indicator of PH and could indicate a life-threatening state (Kondo et al., 2019).

Pathophysiology

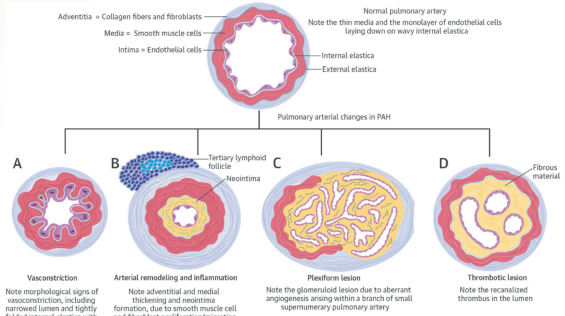
- Group 1 PH is known as pulmonary arterial hypertension (PAH) (Bandyopadhyay et al., 2021).
- In addition to elevated mPAPs, pulmonary artery wedge pressures (PAWP) are <15 mmHg and pulmonary vascular resistance (PVR) > 3 Wood Units (WU) (Bandyopadhyay et al., 2021).
- Group 1 patients undergo pulmonary bed remodeling and loss, leading to increased pressure and RV dysfunction with decreased cardiac output (Bandyopadhyay et al., 2021).
- Group 2 PH originates from left-sided heart impairment leading to increased pressure (Bandyopadhyay et al., 2021).
- Left-sided heart failure and valvular disease are common causes of Group 2 PH (Bandyopadhyay et al., 2021).
- Group 3 PH results from chronic lung diseases like COPD, sleep-related breathing disorders, and interstitial lung disease (Bandyopadhyay et al., 2021).
- Group 3 PH originates from decreased vasculature, distensibility, and vessel recruitment (Bandyopadhyay et al., 2021).
- Group 4 PH patients often have a thrombotic event, most commonly a previous pulmonary embolism resulting in vascular remodeling (Bandyopadhyay et al., 2021).
- Group 5 PH has an unclear or multifactorial pathophysiological process from hematologic, metabolic, systemic, and congenital issues (Bandyopadhyay et al., 2021).
- PH has genetic links, specifically to the bone morphogenetic protein receptor-2 (BMPR2) gene involved in cell differentiation and osteogenesis (Gelzinis, 2022).
- PH originates from the underproduction and overproduction of vasoactive mediators like endothelin, nitric oxide, and prostacyclin (Gelzinis, 2022).
- PH also relates to immunity and metabolic dysfunction (Gelzinis, 2022).
- All forms of PH can lead to RV dysfunction (Bandyopadhyay et al., 2021).



(Adult Congenital Heart Association, 2015)

Pathophysiology Significance

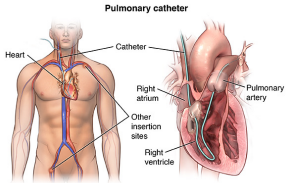
- There are many etiologies for PH, making it important for patients to receive specialized care (Wood et al., 2021).
- Increased lung pressures eventually transmit to the right side of the heart leading to an increase in RV afterload resulting in heart failure (Gelzinis, 2022).
- The RV has a reduced wall density in comparison to the left ventricle, leading to an enhanced response to increased pressures associated with PH (Wood et al., 2021).
- The combined pathophysiological factors leading to RV failure can result in cardiovascular collapse and myocardial ischemia, affecting all organs and bodily functions (Wood et al., 2021).
- Impaired vasoactive mediators like nitric oxide cause impaired coronary perfusion, leading to worsening of myocardial oxygen demands (Wood et al., 2021).
- The pathophysiology of PH leads to impaired glucose handling, mitochondrial differences, and elevated calcium, leading to special considerations for patients (Gelzinis, 2022).
- Due to the complex nature of the PH pathophysiology, a referral to a specialist center is imperative to expedite diagnostic testing and treatment (Mandras et al., 2020).
- Special surgical considerations are necessary due to the pulmonary constriction in patients with PH (Bandyopadhyay et al., 2021).



(Gelzinis, 2022)

Diagnostics

- Early detection of PH is essential with the use of transthoracic echocardiography (TTE) (Barberà et al., 2018).
- Ventilation-perfusion lung scintigraphy for thrombosis detection is pertinent (Barberà et al., 2018).
- Right heart catheterization is important for a baseline hemodynamic status (Barberà et al., 2018).
- Family history, autoimmune testing, echocardiography, and serology testing are important for distinguishing types of PH (Barberà et al., 2018).
- Brain natriuretic peptide (BNP) and *N*-terminal prohormone of BNP are biomarkers for PH (Mandras et al., 2020).



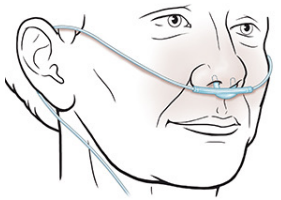
(Cedars Sinai, 2022)

Treatment

- Combination therapy, including a Phosphodiesterase-5 inhibitor and an endothelin-receptor antagonist, is the primary treatment for PH (Kondo et al., 2019).
- Diuretic use for those experiencing fluid accumulation is essential (Barberà et al., 2018).
- Treatment is etiology-dependent (Poch & Mandel, 2021).
- Left heart disease treatment directed towards correction of valvular dysfunction or heart failure diagnosis (Poch & Mandel, 2021).
- Underlying lung disease needs correcting (Poch & Mandel, 2021).
- Pulmonary thromboendarterectomy considered if thrombus involvement in diagnosis (Poch & Mandel, 2021).
- Lung transplant after inadequate treatment (Kondo et al., 2019).

Nursing Implications

- Education is important for patients with PH, especially regarding anticoagulation and treatment options (Barberà et al., 2018).
- The nurse should ensure a thorough assessment with inclusion of history, hemodynamic status, physical condition, and an echocardiogram is completed (Mandras et al., 2020).
- Ensure patient remains normothermic to avoid further pulmonary vasoconstriction (Bandyopadhyay et al., 2021).
- Ensure consistent oxygen saturation monitoring and supplemental oxygen supply to keep saturation >92% (Bandyopadhyay, 2021).
- Maintain low-anxiety environment to minimize pressure changes (Bandyopadhyay et al., 2021).
- Ensure adequate analgesia through use of pain medication and relaxation techniques (Bandyopadhyay et al., 2021).
- Closely monitor oxygenation and ventilation through frequent carbon dioxide and oxygen monitoring (Bandyopadhyay et al., 2021).
- For nurse anesthetists specifically, appropriate anesthesia induction through adequate pre-oxygenation and positive-end-expiratory pressure can prevent adverse events (Bandyopadhyay et al., 2021).
- Nurse anesthetists should consider use of inotropic agents like dobutamine and milrinone to supplement low cardiac output and RV failure (Bandyopadhyay et al., 2021).



(Spectrum Health Lakeland Ear, Nose, and Throat, 2022)

Conclusions

- PH is a chronic, progressive disease that can be difficult and lengthy to diagnose (Bandyopadhyay et al., 2021; Poch & Mandel, 2021).
- A significant consequence of PH is RV dysfunction, leading to severe morbidity and possible mortality (Bandyopadhyay et al., 2021; Steppan & Heerdt, 2021).
- Understanding PH, including the pathophysiology, implications, and presentation, is essential for safe delivery of care (Wood et al., 2021).
- Understanding and recognizing signs and symptoms could lead to early diagnosis and treatment, leading to improvement of outcomes (Kim & George, 2019).
- Specialized care is pertinent for patients with PH to receive tailored care and best outcomes due to the complexity of the pathophysiology behind the disease (Mandras et al., 2020).
- Despite the influx of information regarding PH in recent years, there is still much to know and understand regarding the disease and its implications (Bandyopadhyay et al., 2021).

References

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