Chapter 40

Idiopathic Pulmonary Hypertension

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Case Scenario

A 10-year-old female with newly diagnosed idiopathic pulmonary hypertension is scheduled for placement of a peripherally inserted central catheter in Interventional Radiology. Two weeks prior she presented to the emergency department after a syncopal episode, and her parents reported a history of progressive dyspnea on exertion with even minimal activity. Her SpO_2 on room air was 89%.

Transthoracic echocardiography at that time revealed:

- *Right ventricular hypertrophy with suprasystemic right ventricular pressures*
- *Mild right ventricular dysfunction*

Cardiac catheterization the following day confirmed the diagnosis of pulmonary hypertension, demonstrating pulmonary artery pressures 20 mm Hg greater than systemic, a pulmonary vascular resistance of 15 indexed Wood units and no response to nitric oxide administration. She was started on oral bosentan and sildenafil, intravenous treprostinil, and oxygen via nasal cannula. She remained in the intensive care unit as her medications were titrated to effective doses. She now requires long-term central access for continuous intravenous treprostinil therapy prior to discharge home. She has extreme anxiety and is very fearful about the proposed procedure.

Key Objectives

- Discuss clinical signs and symptoms leading to a diagnosis of idiopathic pulmonary hypertension.
- Describe the classes of medications available for treatment of these patients.
- Discuss anesthetic risk for these patients and discussion of risk with the family.
- Describe perioperative care and airway management strategies for this patient.

Pathophysiology

What is the definition of pulmonary hypertension? How does it differ from pulmonary arterial hypertension?

According to the Paediatric Task Force of the 6th World Symposium on Pulmonary Hypertension in 2018, pulmonary hypertension is defined in adults and children as a mean pulmonary artery pressure (mPAP) >20 mm Hg at rest. Normal mPAP at rest is 15 mm Hg.

Pulmonary arterial hypertension (PAH) is a subset of PH and is defined as PH due to pulmonary vascular disease, with elevated pulmonary vascular resistance (PVR) >3 indexed Wood units (iWU or WU/m^2) alongside a normal pulmonary capillary wedge pressure (PCWP) <15 mm Hg.

Clinical Pearl

Pulmonary arterial hypertension is a subset of PH due to pulmonary vascular disease and is defined by a mean PAP >20 mm Hg at rest and PVR >3 iWU with normal PCWP.

What is the pathophysiology of PAH?

Pulmonary arterial hypertension is a structural pulmonary vascular disease in which smooth muscle cell proliferation and endothelial cell dysfunction lead to a progressive narrowing of the pulmonary vasculature. Additionally, patients with PAH have an imbalance of vasoactive mediators. The production of normally abundant vasodilators, such as nitric oxide and prostacyclin, is decreased, while pulmonary vasoconstrictors, such as endothelin-1 and serotonin, are increased. Both processes combine to cause intravascular thrombosis, vascular remodeling, and eventual destruction of the pulmonary arterioles leading to pulmonary precapillary restriction to blood flow and an increase in the PVR. Initially, vasoconstriction is the major contributor and the changes are reversible. However, as the remodeling continues, the vessels reach an

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