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Portopulmonary Hypertension in Liver Transplantation: Diagnostic Challenges and Perioperative Management of the Right Ventricular Dysfunction

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Abstract

Portopulmonary Hypertension (PPH), a pulmonary vascular complication associated with portal hypertension, affects 2-5% of chronic liver disease patients. PPH's pathogenesis involves pulmonary vasoconstriction, thrombosis, and endothelial and smooth muscle proliferation. In Liver Transplantation (LT) candidates, Right Ventricular (RV) dysfunction arises as the right heart is challenged by high pulmonary pressures, increasing perioperative risks. Mild to moderate PPH is generally tolerated; however, severe cases elevate perioperative morbidity and mortality, especially due to RV failure during reperfusion. Diagnostic approaches, including echocardiography, often underestimate PPH severity, necessitating Right Heart Catheterization (RHC) to confirm diagnosis, particularly in borderline cases. Despite advancements, preoperative screening remains challenging, and intraoperative hemodynamic management is critical. This review discusses PPH's diagnostic complexities and perioperative management strategies in LT, emphasizing early detection, hemodynamic monitoring, and individualized therapy. Effective management of PPH in LT candidates requires a multidisciplinary approach to reduce perioperative complications and optimize patient outcomes.

Keywords: Anesthesia; Echocardiography; Liver transplantation; Portopulmonary hypertension; Right ventricle dysfunction.

Introduction

Elevated pressures in the pulmonary vasculature are frequently observed in patients with severe liver disease, ranging from normal to elevated pulmonary vascular resistance. When pulmonary arterial hypertension is worsened by portal hypertension, with or without significant hepatic damage, it is clinically referred to as Portopulmonary Hypertension (PPH). PPH affects 2% to 5% of individuals with chronic liver disease. The mechanisms underlying this condition are not completely understood but are believed to involve increased resistance to pulmonary arterial flow due to vasoconstriction, intravascular thrombosis, and varying degrees of pulmonary endothelial and smooth muscle proliferation. Several vasoactive, proliferative, and angiogenic mediators are thought to be dysregulated in this complex process [1,2]. The right ventricle, primarily designed to handle volume rather than pressure, may hypertrophy over time to accommodate the increased workload when pulmonary artery pressure rises gradually. However, if the changes occur rapidly or if right-ventricular contractility is compromised, right-ventricular failure may ensue [1].

The current diagnostic criteria for PPH include the presence of portal hypertension, a Median Pulmonary Arterial Pressure (mPAP) >25 mmHg at rest, Pulmonary Vascular Resistance (PVR) >240 dynes.s.cm (>3 Wood units), and Pulmonary Capillary Wedge Pressure (PCWP) <15 mmHg, measured through Right-Heart Catheterization (RHC). PPH is classified into mild (mPAP, 25-35 mm Hg), Moderate (mPAP, 36-45 mm Hg), and severe (>45 mm Hg) categories [8]. The Transpulmonary Gradient (TPG), the difference between mPAP and PAOP, is an additional hemodynamic parameter used to evaluate PPH, with a TPG >12

mmHg suggesting elevated PVR [3].

It is crucial to rule out other causes of pulmonary hypertension, such as volume overload, obstructive or restrictive lung disease, left-heart failure, and obstructive sleep apnea. Moreover, in 30% to 50% of patients with end-stage liver disease, normal pulmonary vascular resistance may be observed alongside elevated pulmonary arterial and venous pressures. This phenomenon, common in Liver Transplant (LT) candidates with a hyperdynamic circulatory state, does not necessarily contraindicate transplantation [1].

The clinical presentation of PPH varies, ranging from asymptomatic to severe symptoms such as dyspnea, fatigue, and peripheral edema. Given that up to 60% of patients with PPH may be asymptomatic at diagnosis, it is recommended that all LT candidates be screened for this condition [4]. When mPAP is less than 35 mmHg, studies have shown no increase in perioperative mortality, supporting LT in patients with moderate PPH and preserved cardiac output and Right Ventricular (RV) function. However, moderate to severe PPH is associated with a higher need for vasodilator therapy, prolonged hospital stays and extended mechanical ventilation postoperatively. Although an mPAP >50 mmHg (or >45 mmHg in some centers) is generally considered an absolute contraindication to LT, some experts suggest that unless PPH is severe and accompanied by RV dysfunction, it should not be an absolute contraindication [5].

The aim of this mini review is to examine the diagnostic and perioperative management challenges posed by Portopulmonary Hypertension (PPH) in Liver Transplantation (LT) patients. Given the heightened perioperative risks associated with Right Ventricular (RV) dysfunction in these patients, this review seeks to explore the limitations of current diagnostic methods, including echocardiography and right heart catheterization, in accurately assessing PPH severity. Furthermore, it aims to highlight key management strategies during the intraoperative and reperfusion phases to mitigate the risk of RV failure. By addressing these critical aspects, the review intends to provide guidance on optimizing outcomes for LT candidates with PPH, emphasizing the importance of early diagnosis, comprehensive hemodynamic monitoring, and individualized therapeutic interventions in this high-risk patient population.

Methodology

A comprehensive literature search was conducted in the Pubmed and Scopus databases using the search terms: anesthesia, echocardiography, liver transplantation, portopulmonary hypertension, right ventricle dysfunction. The search included studies published from 2010 and 2024. Original studies, systematic reviews and relevant clinical guidelines were included. Studies in English, French and Spanish were included. Narrative reviews papers were included if they describe clinical presentation, the diagnostic criteria and anesthetic management for PPH. A variety of studies, including quantitative, qualitative, and mixed-method approaches, were considered to explore the challenges in the diagnosis of PPH that could impact the intraoperative period and the limitations of current diagnostic methods, including echocardiography. Papers were excluded if they did not fit into the conceptual framework of the study. Studies with animals were also excluded. Studies considered relevant were evaluated in full. Relevant information was grouped and presented in a narrative form.

Discussion/Conclusion

In patients undergoing liver transplantation with moderate to severe PPH (mPAP>35 mmHg), mortality rates range from 35% to 50%, with most deaths attributed to right heart failure and cardiopulmonary collapse following reperfusion, massive fluid infusion, or blood transfusion [6]. Previous studies have established that mild to moderate PPH is not a contraindication to LT, but these patients are more susceptible to decreased RV contractile reserve, particularly during the reperfusion phase, leading to a drop in RV ejection fraction [6].

Transthoracic Doppler Echocardiography (TDE) plays a crucial role in evaluating symptomatic cirrhotic patients suspected of having PPH and is recommended for screening all LT candidates by the American Association for the Study of Liver Diseases [7]. Although RHC provides the most reliable hemodynamic assessment, screening for PPH is largely based on Doppler echocardiography. RHC is indicated if the Right Ventricular Systolic Pressure (RVSP) exceeds 50 mmHg, or if there is significant RV hypertrophy or dysfunction observed during a routine TTE exam [5].

However, there are important limitations to preoperative diagnostic methods like TDE, particularly in patients with mild or moderate pulmonary hypertension. The Tricuspid Regurgitation (TR) velocity is used to estimate Pulmonary Artery Systolic Pressure (PASP), but the accuracy of absolute PASP values derived from TR Velocity (TRV) is only moderate. Inaccurate Doppler alignment, eccentric jets, or the absence of a clear TR signal may lead to over- or underestimation of PASP. Consequently, in some patients, PPH may only be diagnosed intraoperatively after the placement of a Swan-Ganz catheter, which provides a more precise measurement of pulmonary artery pressure and can detect previously undiagnosed pulmonary hypertension [8].

The Right Ventricle (RV) faces significant challenges during liver transplantation due to increased preload from volume overload and unfavorable changes in the pulmonary vascular system, typical in cirrhotic patients. Hyperdynamic circulation with low systemic vascular resistance, driven by nitric oxide and other inflammatory mediators, constantly strains the RV, alongside poor cardiac contractility often seen in liver disease. During the anhepatic phase, cross-clamping of the inferior vena cava, portal vein, and hepatic artery dramatically reduces cardiac output. Right ventricular dysfunction, even in a mild HPP may occur [8].

During reperfusion, even patients with mild pulmonary hypertension can experience severe complications due to the sudden increase in venous return and RV overload. Reperfusion syndrome, which affects approximately 30% of LT recipients, can cause a rapid rise in pulmonary arterial pressure, potentially leading to acute RV failure, graft congestion, and subsequent graft failure. Therefore, managing the timing of reperfusion is crucial in these cases, with various interventions such as ECMO, milrinone, intravenous prostacyclin, and inhaled nitric oxide being employed to mitigate these risks [9].

PPH presents significant challenges in the perioperative management of liver transplant patients. Early diagnosis is essential, yet preoperative screening methods like echocardiography have limitations, particularly in detecting mild to moderate PPH. Right heart catheterization remains the gold standard for diagnosis but is often underutilized in patients with mild symptoms. The intraoperative period, particularly during reperfusion,

poses the greatest risk for right ventricular failure, even in cases of mild PPH. Therefore, timely identification and appropriate interventions are critical to improving outcomes. Standardizing pretransplant pulmonary evaluation protocols and optimizing perioperative management are essential steps toward reducing morbidity and mortality in this vulnerable patient population.

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