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REVIEW

Clinical aspects of portopulmonary hypertension

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Summary
Portopulmonary hypertension (PoPH) is an often neglected form of pulmonary hypertension where pulmonary hypertension occurs in the presence of portal hypertension. PoPH is important to diagnose and treat as it may improve the patient's quality of life and improve the outcome after liver transplantation. In this review, we discuss the clinical aspects of PoPH including its pathophysiology, diagnosis, treatment, and prognosis.

Contents

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Introduction

Portopulmonary hypertension (PoPH) is a well-recognized complication of portal hypertension (PH). PoPH has a great impact on quality of life, survival, and suitability for liver transplantation. The aim of this article is to provide a detailed discussion concerning the epidemiology, diagnosis, treatment and clinical significance of PoPH.

Historical background

In 1951, Mantz and Craige reported a peculiar case of a 53-year old woman who expired while undergoing a thoracotomy for hematemesis [1]. The authors noted that the patient's pulmonary artery was enlarged and exhibited forceful pulsations more characteristic of the aorta than of the typically low-pressure pulmonary trunk. The intraoperative findings and subsequent autopsy revealed a stenotic portal vein, a portocaval shunt, and esophageal varices. The pulmonary arteries demonstrated intimal thickening, endothelial proliferation and thrombotic changes. In retrospect, this patient suffered from what subsequently became known as portopulmonary hypertension-a syndrome characterized by pulmonary hypertension in the setting of portal hypertension.

On the basis of this initial description, it was unclear if the association of portal hypertension and pulmonary hypertension was coincidental. However, subsequent reports of similar cases suggested that there was an undeniable relationship between the two conditions. Initially, the observation that PoPH was a distinct clinical entity did not have major practical implications because treatment for pulmonary hypertension and advanced liver disease was not yet available. However, today, in the era of pulmonary vasodilators and orthotopic liver transplantation, the diagnosis of PoPH is imperative as current treatment may improve symptoms, function, and survival.

Definition

The World Health Organization (WHO) classifies PoPH as a form of Group 1 (pulmonary arterial hypertension): mean pulmonary arterial pressure (mPAP) $>$ 25 mmHg and pulmonary artery occlusive pressure (PaOP) $<$ 15 mmHg at rest.

PoPH is defined as pulmonary arterial hypertension associated with portal hypertension. The traditional WHO definition of pulmonary arterial hypertension does not require that the pulmonary vascular resistance (PVR) exceed any particular value. This is because in the setting of a normal or decreased cardiac output, an increased mPAP implies an increased PVR. However, patients with liver disease often develop an abnormally high cardiac output that can result in an elevated pulmonary artery pressure without a concomitant increase in the PVR because of the phenomena of pulmonary vascular recruitment and distention.

Some authors have questioned if the diagnosis of PoPH can be made in the absence of an increased PVR and have labeled this phenomenon as a "hyperdynamic state" [2]. In 2004, the European Respiratory Society (ERS) guidelines required that the PVR exceed 240 dyn s cm^{-5} for the diagnosis of PoPH [3]. The guidelines acknowledged that this PVR cut-off value was arbitrary. PoPH patients who have a PVR $>$240 dyn s cm^{-5} clearly exhibit intimal thickening, smooth muscle hypertrophy and plexiform lesions in the pulmonary vasculature similar to other forms of PAH. Plexiform changes have not been documented in patients

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