Section: General Medicine

Original Research Article

CORRELATION OF PULMONARY ARTERIAL HYPERTENSION WITH SEVERITY SCORES AND CLINICAL PROFILE IN CIRRHOSIS OF THE LIVER

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ABSTRACT

Background: Pulmonary arterial hypertension (PAH) is a relevant complication of liver cirrhosis with adverse implications for prognosis and liver transplantation outcome. While its epidemiology has been characterized, little is known about its relationship to severity scores or clinical characteristics.
Objective: To evaluate the association of PAH with the severity of liver disease (assessed using Child-Pugh and Model for End-Stage Liver Disease (MELD) scores) and the clinical profile of the case patients with liver cirrhosis.
Materials and Methods: This was a two-year cross-sectional study of 50 liver cirrhosis patients. All the patients had clinical examination, haematological and serological investigations, an abdominal ultrasonogram, endoscopy, and transthoracic 2D echocardiography. PAH was considered present if the PASP at rest exceeded 25 mmHg. The level of cirrhosis was based on both Child-Pugh and MELD scores.
Results: Of 50 patients (44 men, six women; mean age 47 years, range 27–66 years), 11 (22%) were diagnosed as having PAH. Dyspnoea was a symptom in 98% of patients and was more frequent, severe, and associated with fatigue in patients with PAH. Female patients had higher proportions of prevalence, and with a shorter duration of alcohol use. PAH strongly correlated with both severity scores: 0% in Child-Pugh A, 11.1% in B, and 36% in C ($p<0.05$); and 0% in MELD ≤10, 13.6% in MELD 11-19, and 50% in MELD ≥20 ($p<0.05$).
Conclusions: Cirrhosis-associated pulmonary arterial hypertension is strongly related to Child-Pugh and MELD scores and to severe clinical conditions. Screening with echocardiography in advanced cirrhosis may help to identify subjects earlier and improve risk-stratification and transplant listing status.
Keywords: Pulmonary arterial hypertension, cirrhosis, severity scores, Child-Pugh class, MELD, clinical profile.

INTRODUCTION

Cirrhosis, the terminal phase of a wide range of persistent liver conditions, continues to be a significant contributor to morbidity and mortality on a global scale.[1] The progression of fibrosis, the development of regenerative nodules, and the alteration of intrahepatic blood vessels result in portal hypertension and liver dysfunction. Nevertheless, the clinical implications of cirrhosis go beyond liver-related issues.[2] Systemic and non-liver-related problems, specifically those affecting the Cardiopulmonary system, significantly impact symptoms, quality of life, and survival. Pulmonary vascular involvement is now acknowledged as an underestimated and clinically valuable aspect of cirrhosis.[3]

The liver-lung axis comprises hepatopulmonary syndrome (HPS) and portopulmonary hypertension (POPH).[4] Hepatopulmonary syndrome presents a condition in which intrapulmonary vasodilatation leads to gas exchange derangements.[5] POPH is a term used to describe pulmonary arterial hypertension associated with portal hypertension.[6] In those cirrhotic patients, PAH is characterised by the persistence of elevated pulmonary arterial pressure and elevated pulmonary vascular resistance (PVR), which can induce right heart strain and

subsequent failure.[7] Late-onset HPS secondary to hyperdynamic circulation is likely to be associated with changes in pulmonary circulation.[8] Variables such as vascular tone, increased shear stress, and remodelling in pulmonary circulation are influenced by the background of response of pulmonary blood vessels to portal hypertension and systemic hyperdynamic circulation.[9]

From a clinical perspective, PAH in cirrhosis is not just theoretical. Patients may also be symptomatic, including dyspnoea on exertion, fatigue, and, rarely, syncope and, when advanced, evidence of right heart failure.[10] These symptoms are nonspecific but can also be caused by manifestations of advanced liver disease (ascites, anemia, deconditioning, or sarcopenia), so that PAH is often subclinical and potentially neglected. PAH is also decremented prognostically with lower functional capacity and a higher risk of decompensation and mortality.[11] The existence of PAH in liver transplant candidates can carry profound implications for preoperative risk identification and management.[12] Despite the repercussions mentioned, standardized testing for PAH is not universally incorporated into cirrhosis management protocols worldwide. Even though right heart catheterization (RHC) remains the gold standard, its invasiveness precludes it from being used as the initial diagnostic tool.[13] Transthoracic echocardiography, however, serves as a valuable non-invasive method for estimating pulmonary pressures and pinpointing patients who require further evaluation.[14] Integrating echocardiography into the standard care for cirrhosis, alongside other clinical and endoscopic assessments in the setting of portal hypertension and transplant assessment, is relatively straightforward.

An additional consideration is the link between PAH and the severity of liver disease itself. Parameters such as the Child-Pugh grade (Classes A–C) and the MELD score are crucial in clinical decision-making, prioritization, and counselling.[15] Should PAH demonstrate associations with these established parameters, it would reinforce the importance of targeted screening in high-risk individuals, aid in prognosis, and potentially influence the timing of referral and optimization for transplantation. Beyond these scores, the clinical presentation, including symptomatology and examination findings, can also provide valuable cues to clinicians for suspecting PAH in the early stages of cirrhosis.[16]

Nevertheless, in everyday clinical practice, the systematic investigation of the correlation between PAH and severity scores and clinical phenotypes is lacking, leading to variable detection rates and delays in referral.[17] There is a practical need for such data to inform decisions on the selection and timing of screening and the interpretation of results within the context of cirrhosis progression. The current study evaluated the association between PAH and Child-Pugh class and the MELD score and clinical characteristics in patients with cirrhotic liver disease. By comparing echocardiographic findings indicating PAH with established markers of severity and symptoms, we aim to identify high-risk groups, facilitate early detection, and offer insights for a practical screening approach that can be incorporated into routine clinical practice and transplant assessment.

MATERIALS AND METHODS

Study design and setting

This cross-sectional, hospital-based study was conducted over two years (September 2017 to August 2019) in the Department of General Medicine at a tertiary care teaching hospital. The study was approved by the institutional ethics committee, and written informed consent was obtained from all participants.

Study population

The study included **50 consecutive patients** with cirrhosis of the liver admitted during the study period. The diagnosis of cirrhosis was established based on a combination of clinical features, biochemical investigations, ultrasonographic evidence of a shrunken or nodular liver with signs of portal hypertension, and endoscopic findings of varices.

Inclusion and exclusion criteria

Patients aged 18 years or older with confirmed cirrhosis of the liver were eligible for inclusion. Exclusion criteria were applied to minimize confounding conditions that might independently influence pulmonary artery pressures. These included: primary cardiac disease, primary pulmonary disease, systemic hypertension, tension ascites, severe anemia (hemoglobin <8 g/dL), chronic pulmonary thromboembolism, and HIV infection. Patients fulfilling any of these exclusion criteria were not enrolled.

Clinical evaluation

All enrolled patients underwent detailed history-taking and thorough physical examination. Particular attention was given to standard clinical features of cirrhosis and PAH, such as abdominal distension, pedal edema, jaundice, exertional breathlessness, fatigue, confusion, and features suggestive of hepatic encephalopathy. On examination, signs of portal hypertension, including splenomegaly, ascites, dilated abdominal veins, spider naevi, palmar erythema, gynecomastia, and other stigmata of chronic liver disease, were carefully documented.

Laboratory and imaging investigations

The baseline laboratory workup consisted of complete blood count, liver and renal profile, coagulation profile (prothrombin time (PT), activated partial thromboplastin time (APTT), and international normalised ratio (INR)), and viral serology testing for hepatitis B surface antigen (HBsAg) and anti-HCV antibodies. Liver morphologic structure, splenic volume, ascites, and portal vein diameter were demonstrated with abdominal ultrasonography. Esophageal varices and other signs of portal hypertension were recorded

during an upper GI endoscopy. Chest x-ray and 12-lead electrocardiography (ECG) were performed in all patients to rule out primary cardiopulmonary disease.

Echocardiographic evaluation

All patients underwent **transthoracic two-dimensional echocardiography (2D ECHO)**. Pulmonary artery systolic pressure (PASP) was estimated by measuring the peak velocity of the tricuspid regurgitant jet and applying the simplified Bernoulli equation. A PASP >25 mmHg at rest was considered diagnostic of pulmonary arterial hypertension. In addition, right atrial and right ventricular dimensions were noted to assess cardiac structural changes associated with elevated pulmonary pressures.

Assessment of the severity of liver disease

The severity of cirrhosis was graded using two widely accepted scoring systems:

1. **Child–Pugh classification** categorizes patients into Class A, B, or C based on clinical (ascites, encephalopathy) and biochemical parameters (bilirubin, albumin, and prothrombin time).
2. **The Model for End-Stage Liver Disease (MELD) score** provides a numerical estimate of liver disease severity using serum bilirubin, serum creatinine, and INR.

Statistical Analysis: Data were compiled and analyzed using descriptive and inferential statistical methods. Continuous variables were expressed as mean ± standard deviation (SD), and categorical variables were presented as frequencies and percentages. The Chi-square test was used to compare categorical variables, while correlation analysis was performed to examine the relationship between pulmonary arterial hypertension, Child–Pugh class, and MELD score. A p-value of <0.05 was considered statistically significant.

RESULTS

Baseline characteristics

The study included 50 liver cirrhosis patients, comprising 44 men (88%) and six women (12%). The mean age was 47 years, ranging from 27 to 66 years. Chronic alcohol intake was the underlying etiology in all cases.

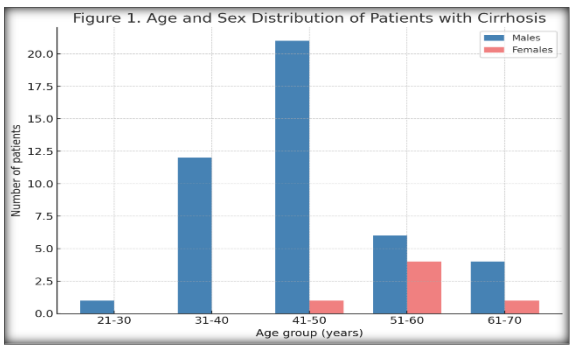

Figure 1: Age and sex distribution of patients with Cirrhosis

Prevalence of pulmonary arterial hypertension

Pulmonary arterial hypertension (PAH), defined as pulmonary artery systolic pressure >25 mmHg, was identified in 11 patients (22%). Although women represented a small proportion of the study population, they showed a higher relative prevalence of PAH than men. The mean age at detection was 46 years in males and 55 years in females.

Clinical profile and PAH

Dyspnoea was the most common symptom, reported in 98% of patients overall, but it was more frequent and severe among those with PAH. Fatigue and exertional intolerance were also more prominent in the PAH group, while syncope was observed occasionally. Classical stigmata of cirrhosis—such as ascites, pedal edema, and jaundice—were universally present, but they did not differ significantly between groups with and without PAH.

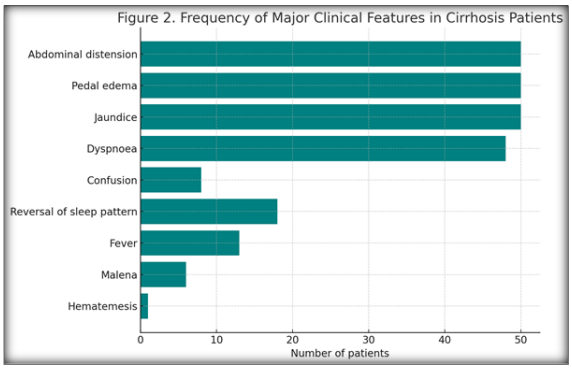

Figure 2: Frequency of major clinical features in Cirrhosis patients

Correlation with Child–Pugh class

The frequency of PAH increased significantly with worsening Child–Pugh stage. None of the patients in Class A had PAH, while 2 of 18 (11.1%) in Class B and 9 of 25 (36%) in Class C were affected. This association was statistically significant (p<0.05).

Correlation with MELD score

A similar trend was observed with MELD scores. Patients with lower MELD (≤10) had no cases of PAH, while prevalence rose to 13.6% in those with MELD 11–19 and reached 50% in those with MELD ≥20. The correlation between PAH and MELD score was statistically significant (p<0.05).

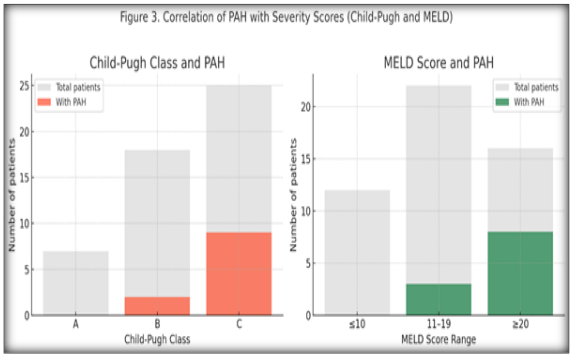

Figure 3: Correlation of PAH with severity scores (Child-Pugh and MELD)

Electrocardiographic findings

Electrocardiographic changes such as right atrial enlargement, right ventricular hypertrophy, and right bundle branch block were observed more frequently among patients with PAH. Although not universal, these findings supported the echocardiographic diagnosis of pulmonary hypertension.

Table 1: Distribution of patients with and without PAH

ECHO finding	Number of patients	Percentage (%)
Patients with PAH	11	22
Patients without PAH	39	78

Table 2: Correlation of PAH with Child–Pugh class

Child–Pugh Class	Total patients	Patients with PAH	Prevalence (%)
A	7	0	0.0
B	18	2	11.1
C	25	9	36.0

Table 3: Correlation of PAH with MELD score

MELD score range	Total patients	Patients with PAH	Prevalence (%)
≤10	12	0	0.0
11–19	22	3	13.6
≥20	16	8	50.0

DISCUSSION

This study highlights the significant association between pulmonary arterial hypertension and advanced liver cirrhosis. Among the 50 patients studied, PAH was detected in 22%, consistent with reported prevalence rates of 15–23% in previous studies on cirrhotic populations.[18] This confirms that PAH, although underdiagnosed, is not an uncommon complication and warrants systematic evaluation in clinical practice.[19] A key observation was the correlation between PAH and the severity of liver disease. None of the patients in Child–Pugh Class A demonstrated PAH, whereas prevalence increased to 11.1% in Class B and 36% in Class C. Similarly, PAH prevalence escalated with higher MELD scores, from 0% in patients with MELD ≤10, to 13.6% in those with scores of 11–19, and reaching 50% in patients with MELD ≥20. These findings are statistically significant and suggest that worsening hepatic dysfunction is directly linked to pulmonary vascular involvement. The trend supports earlier studies that have described portal hypertension and advanced liver disease as essential drivers of pulmonary hemodynamic changes.[20]

The clinical profile of patients with PAH in this study reinforces its impact on patient outcomes. Dyspnoea was nearly universal, but patients with PAH experienced greater severity of symptoms and higher levels of fatigue compared with those without PAH. These symptoms are nonspecific and often overlap with manifestations of cirrhosis, such as ascites or anemia, which may explain why PAH remains under-recognized in practice. However, their prominence in patients with echocardiographic evidence of PAH underscores the need for careful clinical assessment combined with echocardiographic screening. The disproportionate prevalence of PAH among female patients, despite a smaller sample size and shorter history of alcohol use, is an intriguing finding. Similar observations have been reported in pulmonary arterial hypertension unrelated to liver disease, where female sex is considered a risk factor.[21] Hormonal influences and genetic predispositions may play a role, but further studies with larger cohorts are required to clarify this relationship in cirrhotic populations.

The pathophysiology of PAH in cirrhosis is multifactorial. Elevated pulmonary pressures likely reflect a combination of increased blood flow due to hyperdynamic circulation, imbalance of vasodilators and vasoconstrictors, and remodelling of the pulmonary vasculature. Endothelin-1, nitric oxide, and other mediators have been implicated, and their altered balance in cirrhosis may drive pulmonary vascular resistance. The association of PAH with advanced disease stages supports the concept that progressive portal hypertension and systemic circulatory changes amplify these abnormalities.[22]

The clinical implications of these findings are substantial. PAH contributes to increased morbidity, poorer quality of life, and higher perioperative risk in liver transplantation. Preoperative detection is crucial because severe, uncontrolled PAH can even contraindicate transplantation due to unacceptably high operative risk. Therefore, integrating echocardiographic screening into the routine work-up of cirrhotic patients, particularly those with Child–Pugh Class B/C or MELD score≥20, is practical and clinically relevant.

This study has limitations that merit consideration. The sample size was relatively small, which may limit generalizability. The diagnosis of PAH was based on echocardiography, whereas right heart catheterization remains the gold standard for definitive diagnosis and hemodynamic assessment. Nonetheless, echocardiography is a widely validated, non-invasive screening tool, and its use in this study reflects real-world applicability. Despite these limitations, the findings strengthen the evidence that PAH is closely linked with cirrhosis severity and has a distinct clinical profile. These results support a

strategy of targeted screening in higher-risk patients and reinforce the role of echocardiography as a first-line investigation for early recognition of PAH in cirrhosis.

CONCLUSION

This study demonstrates that pulmonary arterial hypertension is a significant complication in cirrhosis of the liver, affecting 22% of patients in the present cohort. Its prevalence was strongly correlated with the severity of liver disease, with higher rates observed in Child–Pugh Class C and MELD scores ≥20. Female patients showed a proportionally higher prevalence, and clinical features such as dyspnoea and fatigue were more prominent in those with PAH. These findings confirm that PAH is not only an under-recognized complication but also a marker of advanced hepatic dysfunction.

Routine echocardiographic screening should be incorporated into the evaluation of cirrhotic patients, particularly those with advanced disease and those awaiting liver transplantation. Incorporating pulmonary pressure assessment into pre-transplant protocols may improve risk stratification and optimize management strategies. Greater clinical vigilance is warranted, since symptoms of PAH are often attributed to liver disease itself, leading to delayed recognition. Future multicentric studies with larger populations and invasive hemodynamic confirmation are recommended to validate these associations and refine screening guidelines.

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