Case Report

Hepatopulmonary syndrome

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Abstract

Hepatopulmonary syndrome (HPS) is a severe complication of liver disease characterized by a triad of advanced liver disease, intrapulmonary vascular dilatations, and arterial hypoxaemia. Key clinical signs include platypnoea and orthodeoxia, with liver transplantation being the only definitive treatment. This article presents a case of suspected Hepatopulmonary syndrome evaluated using bubble contrast echocardiogram and integrated the pathophysiology mechanism described in the literature to explain the clinical and echocardiographic findings**.**

Key Words: Hepatopulmonary syndrome; Budd chiris syndrome; Pulmonary angiogenesis; Atrial hypoxaemia

1. Introduction

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Hepatopulmonary syndrome is an important but often under diagnosed cause of hypoxaemia in patients with chronic liver disease. It is defined by the presence of liver dysfunction, abnormal arterial oxygenation and Intrapulmonary vascular dilatation The prevalence of HPS among patients with cirrhosis ranges from 10% to 30%, with higher rate observed in those undergoing evaluation for liver transplantation. Patient frequently present with dyspnoea and hypoxia, that cannot be fully explained by intrinsic cardiac or pulmonary disease. Contrast echocardiogram has emerged as a key diagnostic tool in identifying Intrapulmonary shunting.

2. Case Presentation

A 47-year-old female patient with known history of decompensated chronic liver disease present with progressive breathlessness, portal hypertension with hepatitis C reactive. Clinical findings of abdominal distention, ascites, reduced urine output, clubbing nails and Spider angioma (spider naevus)- a small red spot with thin blood vessels spreading out like spider legs.

Her vital are stable, Chest X ray, ECG and 2D echo shows normal finding, CT Abdomen contrast shows Budd chiris syndrome with liver Cirrhosis. Her Liver function test has increased total bilirubin count of 2.66 mg/ dl. And reduced albumin of 2.9 g/ dL. Despite of supportive management, the patient continued to exhibit oxygen desaturation that

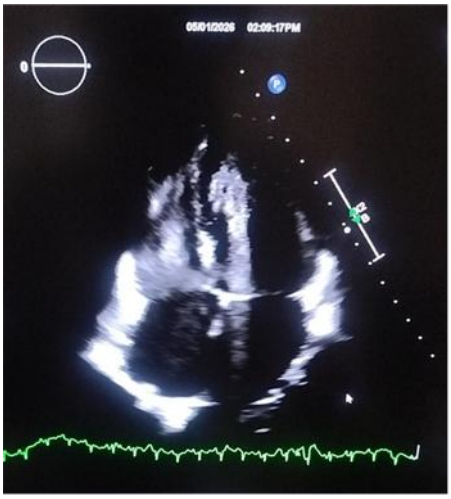
was disproportionate to findings on routine evaluation. Given the unexplained hypoxia in the setting of chronic liver disease a bubble contrast echocardiogram was required to evaluate for a Right to left shunt.

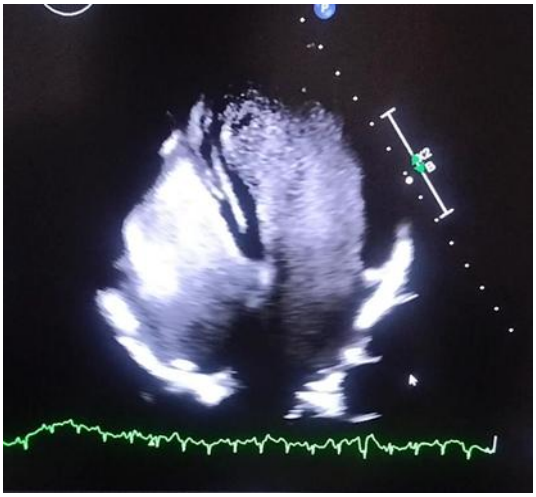
3. Bubble contrast echocardiogram

The primary exposure to bubble contrast echo has been in the evaluation of suspected intracardiac shunt. Though, this case provides a valuable learning opportunity in recognising the diagnostic significant of bubble contrast in patient with complex systemic disease.

The bubble echo was performed using a 10ml agitated saline injected through a peripheral vein. The agitated saline is a combination of 8 ml of normal saline and 2 ml of air. The agitated saline is passed between two 10ml leur lock syringe via a three-way connector. The initial opacification of the right atrium and right ventricle was observed, followed by delayed appearance of microbubbles in the left atrium after 3rd cardiac cycle. The microbubbles transfer within 3 cycle through left atrium refer to as Intracardiac shunt, if there is a delay microbubbles transfer between 4th to 6th cycle is referred as extracardiac or Intrapulmonary shunt. The Intrapulmonary shunt are graded according to the microbubbles, via less than 5 grade I, 5 to 20 microbubbles grade II, more than 20 microbubbles grade III.

However, her bubble contrast echocardiogram shows a positive Intrapulmonary/Extracardiac shunt with grade III Opacification, suggestive of hepatopulmonary syndrome.





4. Pathophysiology of HPS

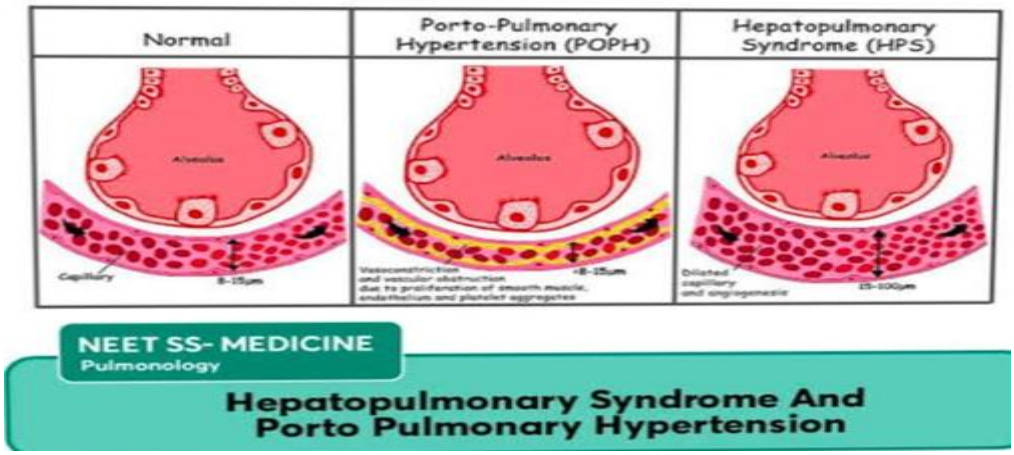
The pathophysiology of HPS is complex and involves abnormal pulmonary vasodilation and angiogenesis within the pulmonary microcirculation. In patient with chronic liver disease, hepatic injury lead to increase production and impaired decrease of vasoactive substance.

Experimental and clinical studies have demonstrated that endothelin 1 plays a central role by stimulating endothelin -B receptors in pulmonary endothelial cells, resulting in regulation of endothelial nitric oxide synthase and increased nitric oxide products.

Excess nitric oxide cause marked pulmonary vasodilation, particularly within the precapillary and capillary vessels. This leads to ventilation -perfusion mismatch a diffusion limitation. As oxygen must travel a greater distance to equilibrate with the red blood

cells. In addition, bacterial translocation from the gut in cirrhotic patient activates pulmonary intravascular macrophages, further increasing nitric oxide synthase. These mechanisms collectively result in functional Right to Left shunt.

Pulmonary angiogenesis has also been implicated in HPS, with studies demonstrate increased vascular endothelial growth factors signaling and capillary proliferation. These structural and functional changes explain the delayed passage of microbubbles through the pulmonary circulation observed on contract echocardiogram



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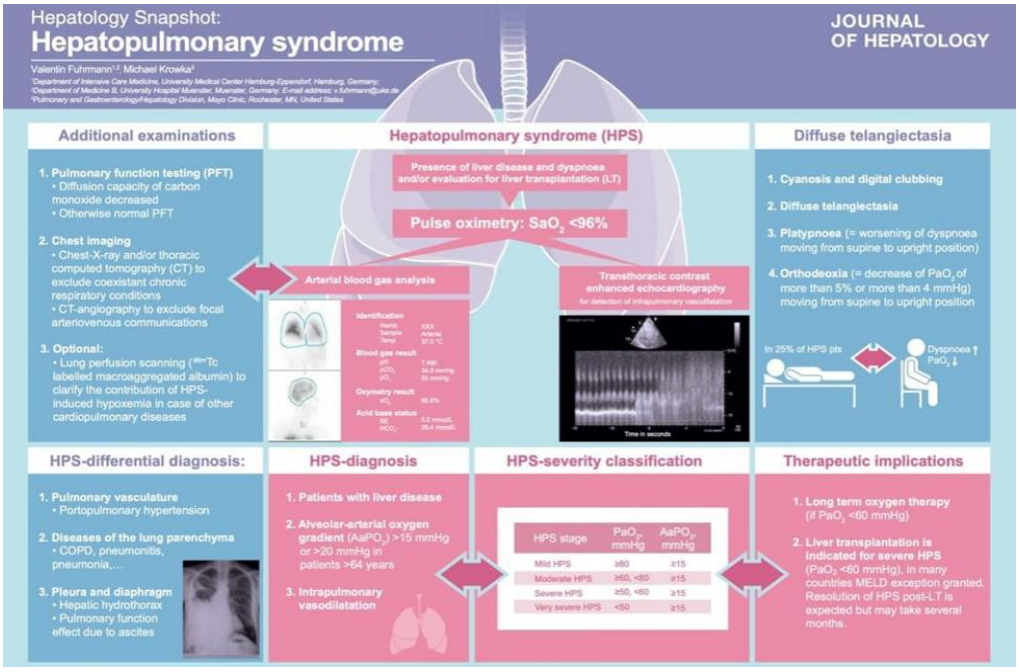
5. Difference between hepatopulmonary syndrome and porto pulmonary hypertension

Hepatopulmonary syndrome (HPS) and portal hypertension are two distinct complications of chronic liver disease that affect different vascular systems and have different clinical and diagnostic implications. Portal hypertension results from increased resistance to blood flow within the cirrhotic liver, leading to elevated pressure in the portal venous system. This manifests clinically as ascites, splenomegaly, and the development of portosystemic collaterals such as esophageal and perisplenic varices. In contrast, Hepatopulmonary syndrome is characterized by abnormal pulmonary vascular dilatation secondary to liver dysfunction, causing intrapulmonary shunting and impaired oxygenation. Patients with HPS typically present with hypoxemia, dyspnea, and characteristic platypnea–orthodeoxia, rather than features of raised portal pressure. From a diagnostic perspective, portal hypertension is identified through imaging and endoscopic findings, while Hepatopulmonary syndrome is confirmed by arterial blood gas analysis and a positive bubble contrast echocardiography showing delayed appearance of microbubbles in the left heart (after 3–6 cardiac cycles), indicating intrapulmonary shunting. Importantly, portal hypertension alone does not produce a positive contrast echo; positivity occurs only when Hepatopulmonary syndrome coexists. Thus, while both conditions arise in the setting of chronic liver disease, portal hypertension represents a portal venous pressure disorder, whereas Hepatopulmonary syndrome is a pulmonary vascular complication with significant implications for oxygenation and transplant prognosis.

Feature	Porto pulmonary hypertension	Hepatopulmonary syndrome
Primary system involved	Portal venous circulation	Pulmonary vascular system
Underlying mechanisms	Increased intrahepatic vascular resistance due to Cirrhosis	Pulmonary capillary dilatation and intrapulmonary shunting due to liver dysfunction
Main haemodynamic changes	Elevated portal venous pressure	Abnormal pulmonary vasodilatation
Typical clinical features	Ascites, splenomegaly, varices	Dyspnea, hypoxemia, platypnea–orthodeoxia
Oxygenation status	Usually normal	Reduced PaO_2 with widened A–a gradient
Cause of symptoms	Venous congestion in portal system	Impaired gas exchange
Role of Porto systemic collateral	Present and prominent	Not responsible for hypoxemia
Bubble contrast echocardiogram	Usually Negative	Positive with delayed bubbles (4–6 cardiac cycles)
Timing of bubbles appearance in LA	No appearance	Delayed appearance
ABG Findings	Usually normal	Hypoxemia
Chest X ray	Usually normal	Ofen normal
Definitive treatment	TIPS, medical therapy, liver transplantation	Liver transplantation (curative)
Prognostic implication	Chronic but manageable	Indicates advanced disease, affects transplant priority



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6. Discussion

HPS is a well recognised pulmonary vascular complications of chronic liver disease and is characterized by the traid of liver dysfunction, atrial hypoxaemia and Intrapulmonary vascular dilatation. In the present case, the patient with decompensated chronic liver disease (child B) and Hepatopulmonary syndrome due to Portalhepato hypertension

Management of HPS is primarily support. Long-term oxygen therapy is recommended to improve symptoms and quality of life in hypoxaemia patient. Optimization of liver disease management includes treatment of underlying Anti HCV infection and prevention of future decompensation. However, pharmacological therapies aimed to reversing the only definitive treatment proven to reverse Intrapulmonary vascular dilatation and improve survival. Early diagnosis using Contrast echo is therefore crucial to facilitate timely referral for transplant assessment. This case highlight the importance of consideration HPS is cirrhotic patient with unexplained hypoxaemia and reinforced the diagnosis and prognostic value of bubble contrast echo in guiding further management.

7. Conclusion

HPS should be suspected in patient with chronic liver disease and disproportionate hypoxia. Bubble contrast echocardiogram is a sensitive, noninvasive diagnostic tool for detecting Intrapulmonary shunting and plays a central role in the evaluation of suspected HPS. Understanding the underlying pathophysiology helps to correlate echocardiographic findings with clinical presentation and support timely referral for definitive management.