CASE REPORT PEER REVIEWED | OPEN ACCESS

Bubble contrast echocardiography study for diagnosing pulmonary arteriovenous shunt in a case of hepatopulmonary syndrome

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ABSTRACT

Introduction: Hepatopulmonary syndrome is an important cause of hypoxemia in children with chronic liver disease. We demonstrate the clinical utility of a simple bedside test to diagnose the condition. Case Report: A 10-year-old with Budd–Chiari malformation presented with severe hypoxemia. Chest X-ray and computed tomography (CT) thorax were normal. Bubble contrast echocardiogram showed return of bubbles to left atrium between the third and fourth cardiac cycle suggestive of pulmonary arteriovenous shunt. Conclusion: Contrast echocardiogram is a sensitive tool to diagnose pulmonary arteriovenous shunt.

Keywords: Bubble contrast echocardiography, Hepatopulmonary syndrome, Pulmonary arteriovenous shunt

INTRODUCTION

Pulmonary arteriovenous shunt is an uncommon cause of hypoxia in children. Pulmonary arteriovenous malformations (PAVM) may be seen as isolated abnormalities or in association with hereditary hemorrhagic telangiectasia. Chronic liver disease is a rare cause of pulmonary arteriovenous shunt in the pediatric age group. It is hypothesized that the lack of some “hepatic factor” may contribute to the development of PAVM in patients with liver disease [1]. We report a case of Budd–Chiari syndrome with severe hypoxia, who was diagnosed to have pulmonary arteriovenous shunt, using bubble contrast echocardiography.

CASE REPORT

A 10-year-old girl, previously diagnosed as Budd–Chiari syndrome with Factor V Leiden mutation, presented with a history of three episodes of hematemesis in last 24 hours. She had undergone variceal ligation and left-sided portal vein stenting one year back and started on anticoagulation but was lost to follow up. In ER, she had severe hypoxia and shock requiring intubation. She was ventilated with following settings: Pressure regulated volume control mode-FiO₂-100%, PEEP-12, R/R-30, TV-120 mL. Chest X-ray was normal. But PaO₂/FiO₂ ratio was <60. The possibility of pulmonary thromboembolism was ruled out by CT thorax angiography and echocardiography. Considering the underlying illness, hypoxia was thought to be due to hepatopulmonary syndrome.

Subsequently, bubble contrast echocardiography was done showing appearance of bubbles in left atrium between the third and fourth cardiac cycle (Video 1 and Figure 1). Diagnosis of pulmonary arteriovenous shunt was confirmed.
secondary to hepatopulmonary syndrome was confirmed with technetium 99 lung perfusion scan showing large shunting at the pulmonary capillary bed level.

In view of intractable liver failure, liver transplantation was planned, but the child developed ventilator-associated pneumonia and sepsis and succumbed to the illness.

**DISCUSSION**

Contrast echocardiography using agitated saline bubbles of minimum of 15 microns can be used as a diagnostic tool for hepatopulmonary syndrome [2]. Normally, microbubbles are trapped in pulmonary vasculature and absorbed. However, in presence of pulmonary arteriovenous shunts like hepatopulmonary syndrome or pulmonary arteriovenous malformations, the bubbles are seen in the left heart after the third heartbeat [3], usually between the third and sixth heartbeat as demonstrated in Video 1. Whereas, in presence of intracardiac right to left shunts, these microbubbles are seen in the left heart within the first three cardiac cycles [4]. Perfusion scans using technetium-labeled albumin macroaggregates (Tc-99m MAA) are diagnostic tests for pulmonary arteriovenous malformations and permit precise shunt quantification [5], but they need mobilization of the patient outside ICU, which is often associated with potential risks in critically ill patients, whereas bubble contrast echocardiography can be done at bedside without any such concerns.

**CONCLUSION**

Contrast echocardiography is a sensitive bedside test to screen for pulmonary arteriovenous shunt.

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**Author Contributions**

Sandip Gupta – Conception of the work, Design of the work, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related
to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Karthik Arigela – Conception of the work, Drafting the work, Final approval of the version to be published. Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Sweta Mohanty – Conception of the work, Design of the work, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published. Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Authors declare no conflict of interest.

Data Availability
All relevant data are within the paper and its Supporting Information files.

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