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Persistent Left Superior Vena Cava (PLSVC) Draining into an Unroofed Coronary Sinus: Successful Trans-catheter Occlusion of PLSVC with an Amplatzer Vascular Plug

By Harshida Chaudhari,¹ MD; Thomas Doyle, MD; Ashish Madhok, MD, FACC; and Ashok Mehta, MD, FACC

ABSTRACT

Persistent left superior vena cava (PLSVC) draining into an unroofed coronary sinus is a rare congenital anomaly. It may go undiagnosed due to lack of specific signs and symptoms. We report a child with this condition who presented with persistent unexplained hypoxemia. The diagnosis was made by contrast echocardiography and was later confirmed by cardiac catheterization and angiography. A trans-catheter occlusion of the left superior vena cava was performed successfully with an Amplatzer vascular plug (St. Jude Medical, Inc., USA). His oxygen saturation returned to normal immediately after the procedure.

INTRODUCTION

Unroofed coronary sinus, a rare form of congenital heart disease, is commonly associated with persistent left superior vena cava (PLSVC) that drains into the coronary sinus.¹ PLSVC results from impaired degeneration of the left anterior cardinal vein.² PLSVC occurs in 0.1 to 0.5 percent of the general population.^{1,2} The majority of the PLSVC drains into the right atrium via coronary sinus without any problems. However, approximately eight to 10 percent of the PLSVC drains into the left atrium, typically through an unroofed coronary sinus, and can be a risk factor for thromboembolism due to right-to-left shunt.^{3,4}

To the best of our knowledge, there are few reported cases of this condition and their management in the literature. Before the advent of echocardiography, the precise diagnosis of this entity was only possible with invasive procedures like angiography during open heart surgery or autopsy.⁵ Today, there are several types of occlusion devices and thrombotic coils are available for the occlusion of large venous vessels. Hares et al., reported the use of the Amplatzer vascular plug in four different children with complex congenital heart disease.⁶ We present a rare case of an apparently normal child who presented with persistent unexplained hypoxemia and was found to have a PLSVC draining into an unroofed coronary sinus. His hypoxemia instantly resolved after deployment of the Amplatzer vascular plug in the PLSVC.

CASE REPORT

A two-year-old Caucasian male was seen in the emergency room for difficulty in breathing. He had a normal physical examination except for minimal wheezing. His oxygen saturation was 88% by finger pulse oximeter in room air. He had a normal chest Roentgenogram. His oxygen saturation improved to 91% after he was given supplemental oxygen and albuterol nebulizer treatment. However, his oxygen saturation in room air remained 88%. He was referred to us for further evaluation. He had no complaints at that time. He had no family history of congenital heart disease, arrhythmia or sudden cardiac death. His oxygen saturation was 88% in room air by pulse oximetry. He was alert, active and without any distress. His physical examination, 12-lead surface electrocardiogram, trans-thoracic two-dimensional echocardiogram, and color Doppler were normal.

He was rescheduled for a trans-thoracic echocardiogram with contrast. Under conscious sedation, contrast echocardiography was performed with 10ml agitated saline administered through a vein in the left forearm. There was the instantaneous filling of the left atrium and left ventricle with contrast (Figure 1), followed by opacification of the right heart a few seconds later (Figure 2). The patient was referred to Vanderbilt University Medical Center in Nashville for cardiac catheterization and possible vascular occlusion of the PLSVC. Under general anesthesia, a 7-French venous sheath was placed in the left jugular vein for intervention reasons and 6-French venous and arterial sheaths were

placed through the right groin. After heparinization, complete right and left heart catheterization was performed. After the initial hemodynamic, angiography was performed in the right and left superior vena cava. Cardiac catheterization findings confirmed the presence of bilateral superior vena cava with PLSVC draining into the left atrium via an unroofed coronary sinus (Figure 3). He had no left brachiocephalic vein, but there were several venous connections between the right and left superior vena cava. It was decided to proceed with the intervention to the PLSVC. Since there was no change in the left jugular vein pressure with temporary occlusion of PLSVC with a 14mm Amplatzer vascular plug, the PLSVC was permanently occluded with the 14mm Amplatzer vascular plug (Figure 4). He tolerated the procedure well, and his saturation improved to 99% with pulse oximeter in room air. He was discharged home on 81mg acetyl salicylic acid a day and was advised for subacute bacterial endocarditis prophylaxis before any procedure. On follow up one week later, his oxygen saturation was 98% in room air with pulse oximetry. A follow up trans-thoracic echocardiogram was normal.

DISCUSSION

Unroofed coronary sinus is a rare form of congenital anomaly. It is morphologically classified into four different types: I, completely unroofed with PLSVC; II, completely unroofed without PLSVC; III, partially unroofed mid-portion; and IV, partially unroofed terminal portion.¹ Unroofed coronary sinus with PLSVC has been found to occur in patients with and without congenital heart diseases.² Contrast echocardiography through the left arm, and cardiac MRI/CT are helpful to diagnose this condition pre-operatively.⁷

PLSVC associated with unroofed coronary sinus can be occluded or ligated during surgery if sufficient collateral channels are present for venous drainage.⁴ The adequacy of the collateral network can be assessed during cardiac catheterization by temporary balloon occlusion of the PLSVC. A persistent increase in mean pressure of greater than 10mm of Hg in proximal veins is considered high risk for the development of cerebral edema or superior vena cava syndrome.⁴ With the advent of the percutaneous closure of PLSVC with a vascular occlusion device, open heart surgery can be avoided in uncomplicated cases. As reported by Hares et al.,⁶ the Amplatzer vascular occlusion device is relatively easy to deploy, safe and well tolerated in children with complex heart disease. Similarly, Moustafa et al.,⁸ reported a successful occlusion of PLSVC with an Amplatzer vascular plug in an adult patient. There are no long-term adverse events reported with the Amplatzer vascular occlusion device.

The treatment of PLSVC with unroofed coronary sinus is still controversial.⁵ Surgical procedures for unroofed coronary sinus vary depending upon associated cardiac anomalies. In an isolated case of unroofed coronary sinus without the left superior vena cava, roofing the unroofed portion or patch closure of the coronary orifice are usually employed. Care is required during the procedure as complications like conduction disturbances including complete atrio-ventricular blocks can occur.⁷ Patients with unroofed coronary sinus and PLSVC are at a higher risk for systemic embolization. An Amplatzer vascular plug or a similar vascular device should be considered over surgery in an uncomplicated case.

CONCLUSION

Increased awareness of this rare condition (unroofed coronary sinus with or without PLSVC) may aid clinicians in early detection and prompt intervention. Contrast echocardiography with injection of contrast through a vein in the left forearm helps to diagnose this condition. Minimally invasive treatment should be considered in the absence of complex congenital heart disease. Trans-catheter occlusion of PLSVC with an Amplatzer vascular plug is relatively easy, safe, and well tolerated by patients. A vascular plug should be considered over surgery in an uncomplicated case.

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Legends

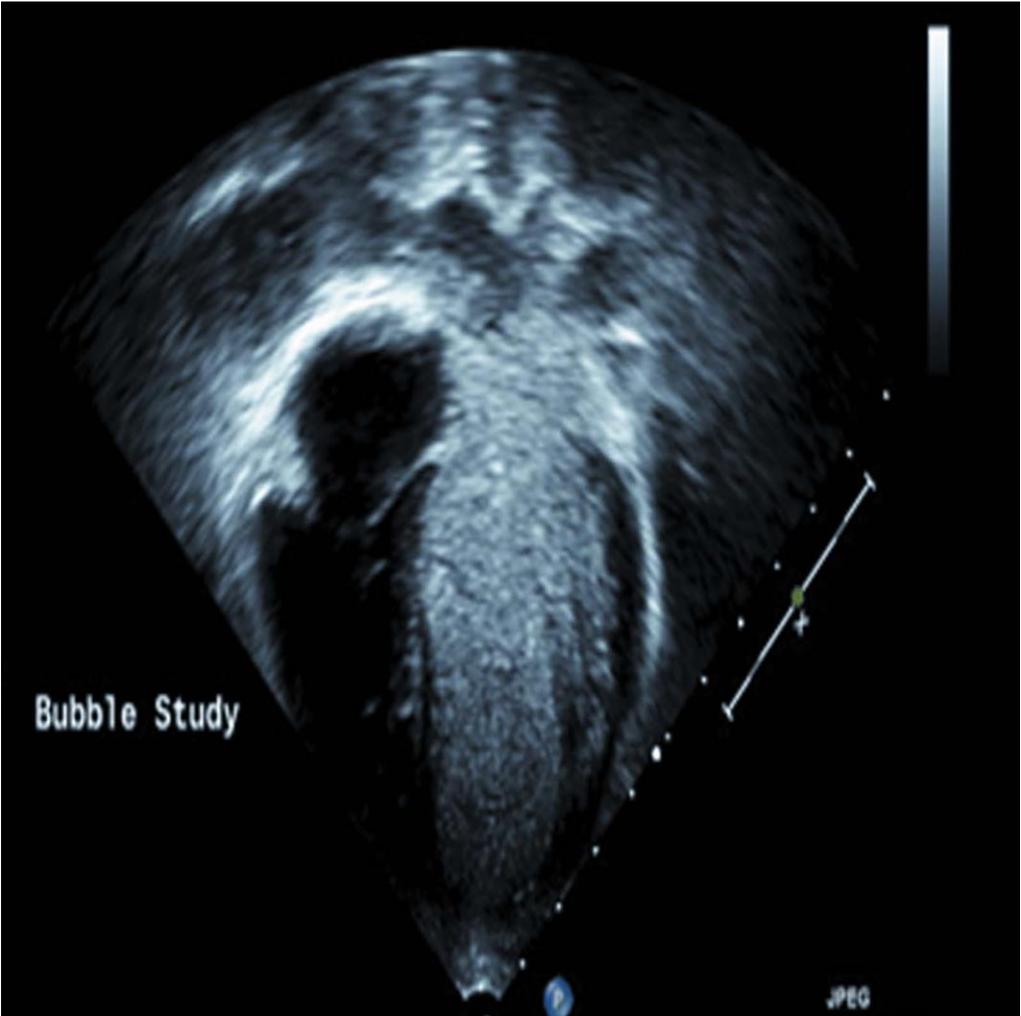


Figure1. Contrast 2-D trans-thoracic echocardiogram shows instant opacification of the left atrium and left ventricle after injection of agitated saline contrast from the left arm.

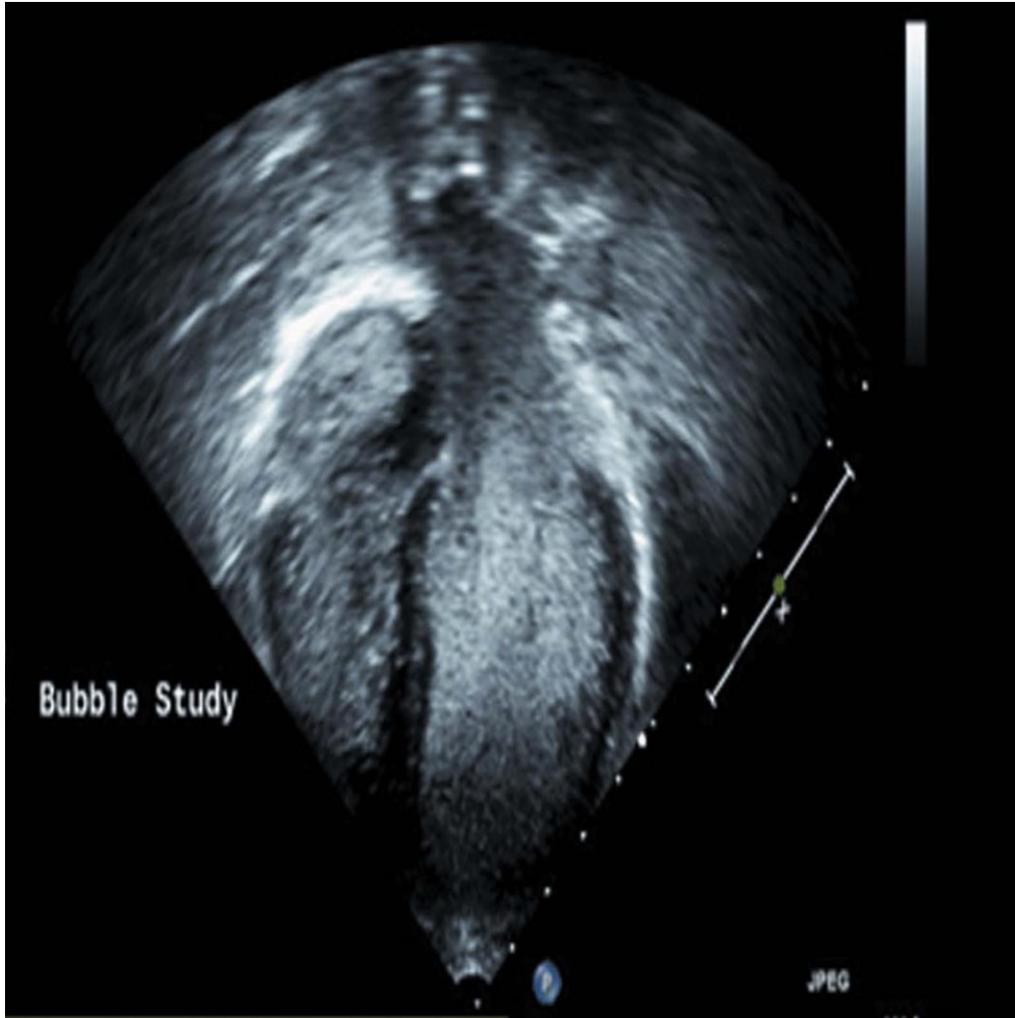


Figure2. Contrast 2-D trans-thoracic echocardiogram shows opacification of the right atrium and ventricle a few seconds later in the same patient.

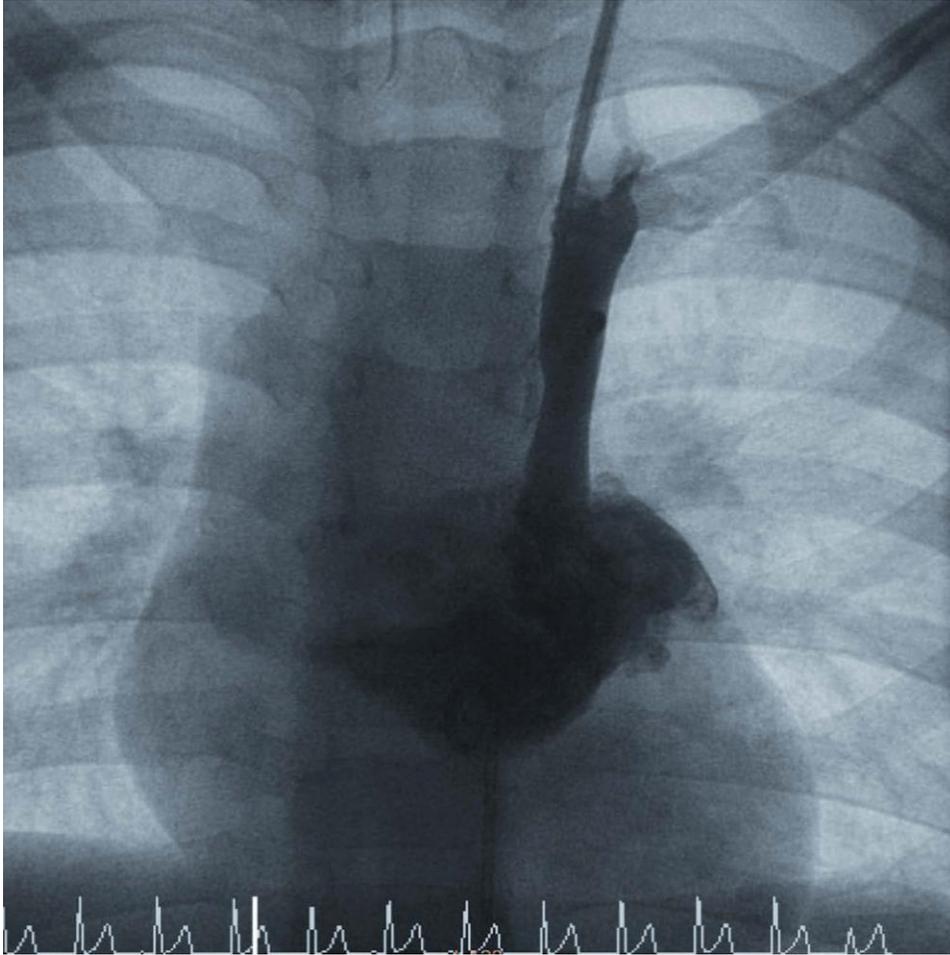


Figure3. Cine-angiogram shows PLSVC draining into an unroofed coronary sinus with opacification of the left atrium.

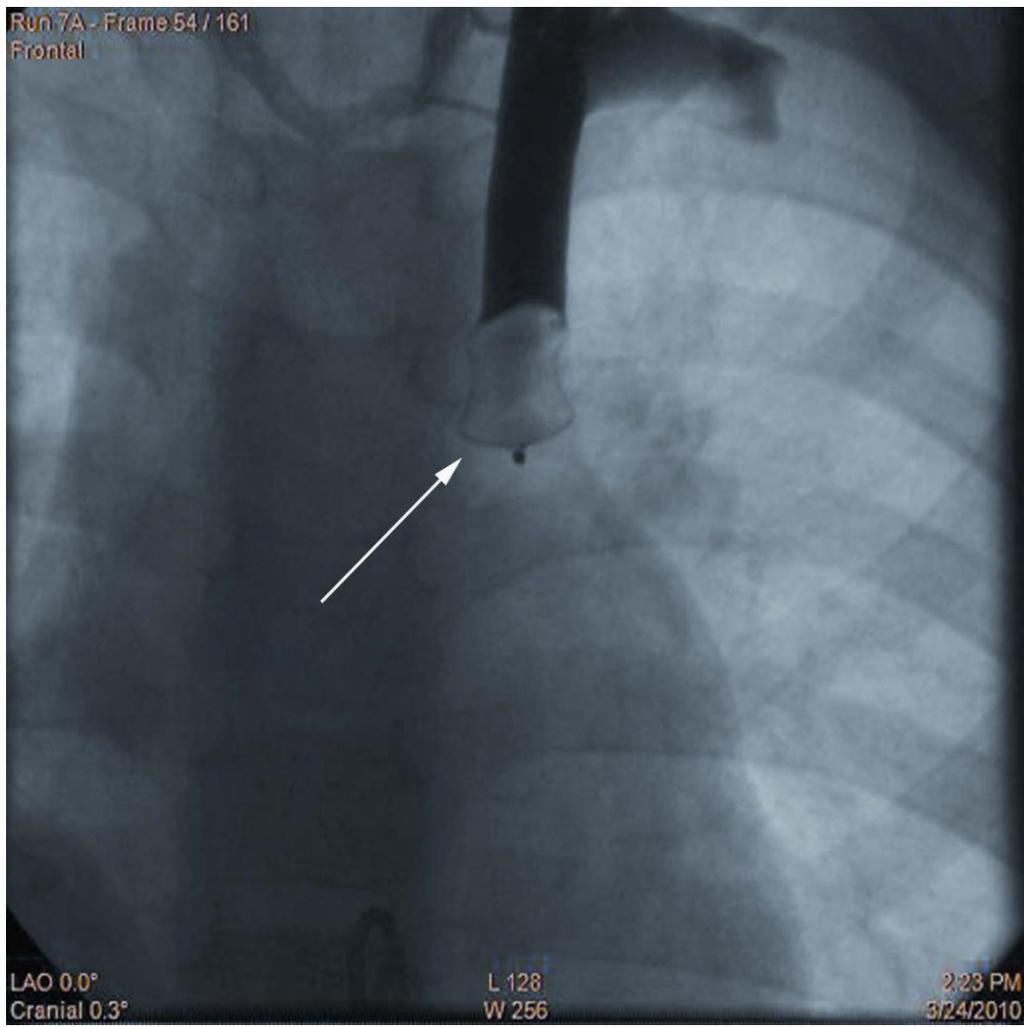


Figure4. Cine-angiogram shows complete occlusion of PLSVC after deploying a 14mm Amplatzer vascular plug (arrow).