# Contrast echocardiogram in diagnosing rare type of persistent left superior vena cava

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**SUMMARY**

**We report a rare case of persistent left superior vena cava (PLSVC) with direct drainage into the left atrium in a 3-years-old boy who had been electively admitted for hypospadias repair, when he was noticed to have finger clubbing and mild hypoxia but was otherwise asymptomatic. The diagnosis of PLSVC can be made without an invasive tool as direct drainage of PLSVC into the left atrium be visualised using transthoracic echocardiography (TTE) by injecting agitated saline into the left arm.**

## INTRODUCTION

The incidence of PLSVC is approximately 0.3 percent in the general population[1] and the prevalence is variable and higher in patients with congenital heart disease.[2] Nearly half of all patients with PLSVC have other cardiac malformations such as atrial septal defects, endocardial cushion defects or Tetralogy of Fallot.[3]

The formation of the embryonic venous system is complex and subject to substantial anatomical variation. During normal development, the anterior cardinal veins, which drain the head, neck, and arm, unite with the posterior cardinal vein in the early embryonic stage and enter the heart as the right and left horn of sinus venosus. Most of the left-sided cardinal system is obliterated, leaving only the coronary sinus and a remnant known as the Ligament of Marshall. Failure of left anterior cardinal vein obliteration gives rise to PLSVC. This usually drains into the right atrium through the coronary sinus.

On rare occasions, when a developmental arrest occurs at an earlier stage, there is an absence of the coronary sinus and the PLSVC drains directly into the left atrium. This is usually asymptomatic and incidentally diagnosed when cardiovascular imaging is performed for unrelated reasons. We report a rare case of PLSVC that drains directly into the left atrium, diagnosed using Transthoracic echocardiography with agitated saline injected intravenously through the left arm.

## CASE PRESENTATION

A 3-year-old boy was electively admitted to the paediatric surgical ward for hypospadias repair. Physical examination revealed a resting hypoxia of 90% on room air with normal respiratory effort and finger clubbing. His electrocardiogram and chest x-ray were normal. There was no lung condition detected to explain patient's condition and the patient was referred to the cardiologist for further evaluation. A TTE was performed and showed the PLSVC draining into the left atrium (Figure 1) with the right superior vena cava draining correctly into the right atrium. No other cardiac abnormality was detected. A follow-up agitated saline ("bubble") study was performed. TTE with agitated saline injected intravenously through the patient's left arm showed a large bolus of bubbles entering into the left atrium first, then immediately into the left ventricle (Figure 2). This finding was confirmed using Contrast-Enhanced Computed Tomography Pulmonary Angiography (CTPA). CTPA showed the left brachiocephalic vein draining into the left atrium via PLSVC. The patient was then referred to the cardiothoracic surgeon for early surgical intervention.

## DISCUSSION

The PLSVC results from failure of regression of the left cardinal vein. PLSVC occurs in several anatomical variations. PLSVC can coexist with the right SVC in up to 80% to 90% of cases. The left innominate vein may be completely absent in 65% of cases. In approximately 80% to 92%, the PLSVC drains into the right atrium through the coronary sinus. This usually causes no haemodynamic disturbances.[4] In approximately 10% to 20%, the PLSVC drains into the left atrium through the unroofed coronary sinus.

This patient falls under the minority subtype group of PLSVC in which the PLSVC drains into the left atrium through the unroofed coronary sinus. As such, he has systemic arterial desaturation which is caused by the mixing of LSVC blood (deoxygenated blood) with pulmonary venous blood (oxygenated blood) in the left atrium. If it is not associated with other congenital cardiac anomalies, then the only clinical manifestation is cyanosis although it can be rarely associated with systemic emboli, stroke[5] and abscesses.

The diagnosis of PLSVC can be confirmed by non-invasive investigations. Echocardiography is the definitive imaging modality in most patients and has good sensitivity and specificity. If the diagnosis is still in doubt, a contrast injection (agitated saline) in a left arm vein establishes the diagnosis by the demonstration of microbubbles in the left atrium before they appear in the right atrium (if patent foramen ovale or interatrial defect present). These findings can be further confirmed by CT with contrast. An alternative investigation is

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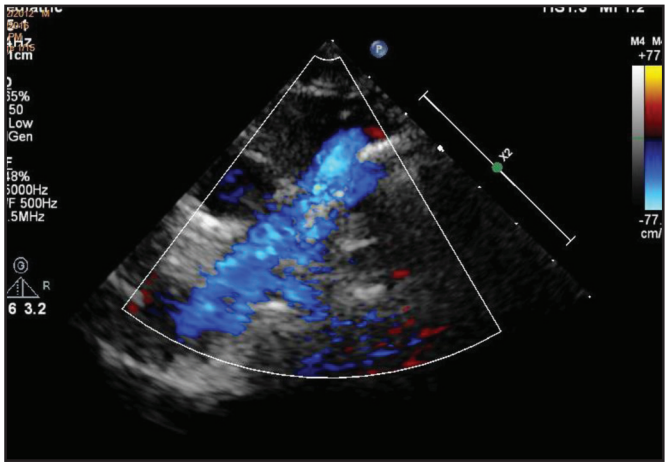

**Fig. 1:** TTE with the suprasternal image showing colour Doppler flow mapping confirming flow from the left superior vena cava to the left atrium.

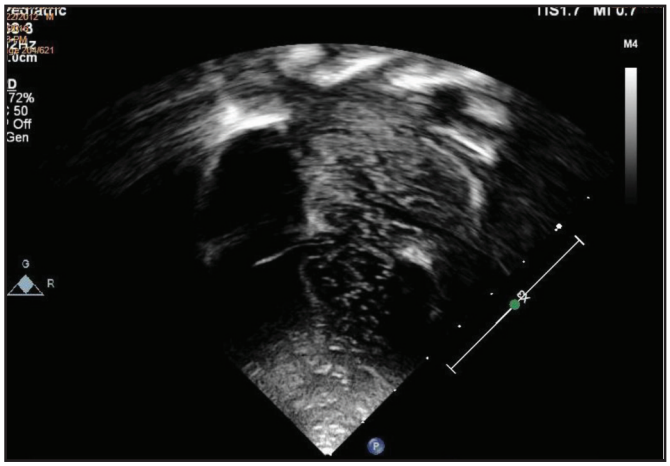

**Fig. 2:** TTE with agitated saline injected intravenously through the left arm. Apical 4-chamber image showing a large bolus of bubbles entering into the left atrium first instead of the right atrium, then into the left ventricle.

MRA which is especially helpful if the anatomy has not been completely delineated.

Cardiac catheterization is unnecessary in most patients with isolated PLSVC. Diagnosis using cardiac catheterization is established by the demonstration of a step-down in oxygen saturation between the pulmonary veins and the LA and by left SVC-selective angiography.

The differential diagnosis for asymptomatic central cyanosis with a normal echocardiogram includes pulmonary arteriovenous shunt or congenital methaemoglobinaemia. The duration of cyanosis as well as symptoms related to cardiovascular and pulmonary systems are important points to establish in the history. The diagnosis of methaemoglobinaemia is further supported by a normal arterial blood gas analysis.

## CONCLUSION

PLSVC with direct drainage into the left atrium is rare and usually asymptomatic. Patients may however, have signs such as unexplained cyanosis and clubbing. Isolated PLSVC can be diagnosed by contrast echocardiography with using proper techniques.

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