

Persistent left superior vena cava – the value of an agitated saline contrast study

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A persistent left superior vena cava (PLSVC) draining to the coronary sinus (CS) is the most common venous anomaly of the thorax, affecting approximately 0.5% - 2% of the general population, and is present in up to 10% of patients with other congenital cardiac anomalies.⁽¹⁾ The embryological development of the thoracic venous system is complex and subject to significant variation. Usually, most of the left cardinal system involutes, leaving only the coronary sinus, which drains the cardiac veins, and the ligament of Marshall (a remnant of the left superior vena cava).⁽²⁾ The presence of a PLSVC is usually an incidental finding at either echocardiography, cardiac catheterisation or device implantation.⁽¹⁾ The typical echocardiographic findings are that of a dilated CS, which can be appreciated on the parasternal long axis view, the parasternal short axis at mitral valve level, the apical 2 chamber view and a modified apical four chamber view scanning down to visualise the CS (Figure 1). A contrast study with agitated saline (“bubble study”) done via the left brachial vein will demonstrate the dilated CS draining into the right atrium (Figure 2).

A rarer anomaly is that of a PLSVC with an unroofed coronary sinus (UCS). There are four types of UCS as classified by Kirklin and Barratt-Boyes:⁽³⁾

- Type I: Completely unroofed, with PLSVC.
- Type II: Completely unroofed, without PLSVC.
- Type III: Partially unroofed in the midportion.
- Type IV: Partially unroofed at the terminal portion.

This can be difficult to diagnose via standard transthoracic echocardiography.^(3,4) The presence of a dilated right heart in a

patient with a PLSVC should raise the suspicion of an associated cardiac abnormality causing a left to right shunt. Common causes would include an atrial septal defect, a sinus venosus defect and anomalous pulmonary venous drainage. The possibility of an UCS must also be considered. This defect may be difficult to demonstrate anatomically. The addition of a contrast study with agitated saline to identify the defect functionally has been shown to increase the accuracy of transthoracic echocardiography in making the diagnosis and delineating the type of UCS, and is currently the initial investigation of choice to diagnose this rare condition (Figure 3).^(5,6) Further studies such as transoesophageal echocardiography, cardiac catheterisation, cardiac computerised tomography and cardiac magnetic resonance imaging are useful when for planning for surgical correction.

The case shown in Figures 1 and 2 was that of a 15-year-old boy presenting with palpitations. The PLSVC was initially



FIGURE 1: Modified apical four chamber view scanning down to visualise the dilated coronary sinus draining into the right atrium. A few bubbles resulting from an intravenous infusion on the left arm can be seen entering the right atrium and right ventricle, both of which are non-dilated.

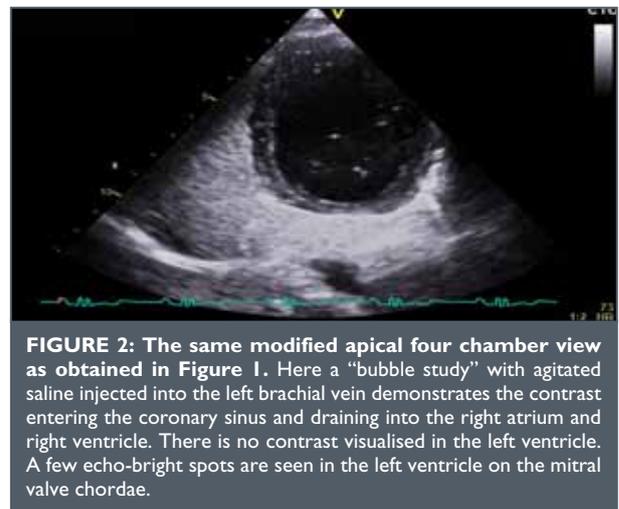


FIGURE 2: The same modified apical four chamber view as obtained in Figure 1. Here a “bubble study” with agitated saline injected into the left brachial vein demonstrates the contrast entering the coronary sinus and draining into the right atrium and right ventricle. There is no contrast visualised in the left ventricle. A few echo-bright spots are seen in the left ventricle on the mitral valve chordae.

misdiagnosed as an abnormality of the posterior leaflet of the mitral valve (based on misinterpreting the roof of the dilated coronary sinus as part of the posterior mitral valve leaflet in the parasternal long-axis view) for which the patient was referred to our valvular heart disease clinic for assessment. He is being followed up by the referring clinician.

The case shown in Figure 3 was that of a 37-year-old female patient referred for echocardiography. She presented with right heart failure and cardiomegaly on chest radiography. A standard transthoracic echocardiogram was done and a diagnosis of a low atrial septal defect (ASD) was made with a left to right shunt. There was associated anomalous hepatic venous drainage. Cardiac catheterisation was performed to determine pulmonary vascular resistance, during which a persistent left superior vena cava (PLSVC) was diagnosed. A contrast study done with agitated saline injected into the patient's left brachial vein confirmed an UCS with contrast entering the left atrium first before crossing the ASD into the right atrium and the mitral valve into the left ventricle (Figure 3). The patient underwent successful surgical correction.

These 2 cases demonstrate the value of an agitated saline contrast or "bubble study" in a patient presenting with a PLSVC, particularly if the right heart is dilated, suggesting an associated

left to right shunt. An UCS can be difficult to demonstrate anatomically and the addition of an agitated saline contrast study enhances the diagnostic accuracy of echocardiography by demonstrating the functional defect. This differentiation is important as an isolated PLSVC requires no treatment whereas an associated UCS may require complex corrective surgery.

Conflict of interest: none declared.

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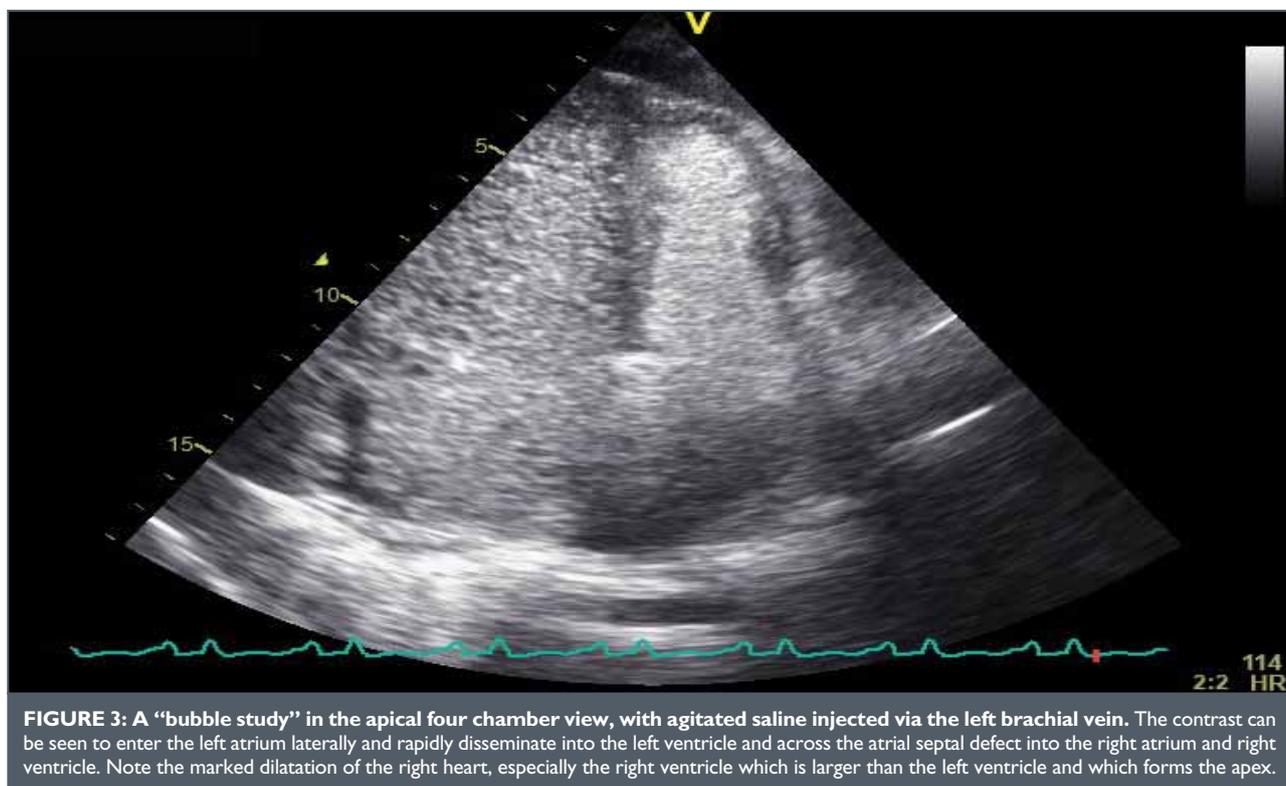


FIGURE 3: A "bubble study" in the apical four chamber view, with agitated saline injected via the left brachial vein. The contrast can be seen to enter the left atrium laterally and rapidly disseminate into the left ventricle and across the atrial septal defect into the right atrium and right ventricle. Note the marked dilatation of the right heart, especially the right ventricle which is larger than the left ventricle and which forms the apex.